AIDS - Children with HIV infection and their families: the Surgeon General's workshop on AIDS*

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The first meeting to discuss pediatric AIDS took place in 1984 and the second in 1986. But, the severity of the situation was not really brought to light until April, 1987, when U.S. Surgeon General C. Everett Koop invited 150 health care providers, researchers, media representatives, and lay people to Children's Hospital of Philadelphia for a conference workshop. Koop charged the group with making recommendations to help set national policy regarding this growing problem. This review is based on the participation of the two pediatric dentists invited to the workshop (coauthors of this manuscript).

Definition

Childhood-acquired immonodeficiency syndrome (AIDS) is defined by the Centers for Disease Control (CDC) for the purposes of public health, epidemiologic studies, disease surveillance, prevention programs, and health care planning and policy (CDC 1984).

Infants and Children with Perinatal Infection up to 15 Months of Age

Infection in infants and children up to 15 months of age who were exposed to infected mothers in the perinatal period may be defined by one or more of the following:

1. The identification of the virus in blood or other tissues; and/or
2. The presence of HIV (human immunodeficiency virus) antibody as indicated by a repeatedly reactive screening test (e.g., enzyme immunoassay) plus a positive confirmatory test (e.g., Western blot, immunofluorescence assay) in an infant or child with abnormal immunologic test results indicating both humoral and cellular immunodeficiency, and meeting the requirements of one or more of the subclasses listed under P-2 (Table 1, next page); and/or
3. Any child who meets the previously published CDC case definition for pediatric AIDS (CDC in press; CDC 1985).

The infection status of other perinatally exposed seropositive children up to 15 months of age who lack the above immunologic and clinical criteria is indeterminate. These infants should be followed for HIV-related illness and tested at regular intervals for persistence of antibody to HIV. Infants and children who: (1) become seronegative; (2) are virus culture negative (if performed); and (3) continue to have no clinical or laboratory abnormalities associated with the HIV infection are unlikely to be infected.

Older Children with Perinatal Infection and Children with Other Modes of Transmission

HIV infection in these children is defined by one or more of the following:

1. The identification of virus in blood or tissues; and/or
2. The presence of HIV antibody (positive screening test plus confirmatory test) regardless of whether immunologic abnormalities or signs or symptoms are present; and/or
3. Any child who meets the previously published CDC case definition for pediatric AIDS (CDC in press; CDC 1985).

These definitions apply to children under 13 years of age. It is recommended that HIV infection in adolescents be classified according to the adult classification system (CDC 1986).

Epidemiology

Data through mid-April, 1987, compiled by the CDC showed the following statistics. Of the 35,397 cases of AIDS in the United States, there were 600 cases of pediatric AIDS that fit the CDC definition in the United States; of these, 471 cases of AIDS were in children under age 13 years and 139 cases in children aged 13-19 years. Sixty per cent of these children have died. An additional

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TABLE 1. Classification of HIV Infection in Children Under 13 Years of Age*

Class P-0. Indeterminate Infection
Class P-1. Asymptomatic Infection
Subclass A. Normal Immune Function
Subclass B. Abnormal Immune Function
Subclass C. Immune Function Not Tested
Class P-2. Symptomatic Infection
Subclass A. Nonspecific Findings
Subclass B. Progressive Neurologic Disease
Subclass C. Lymphoid Interstitial Pneumonitis
Subclass D. Secondary Infectious Diseases
Category D-1. Specified secondary infectious diseases listed in the CDC surveillance definition for AIDS
Category D-2. Recurrent serious bacterial infections
Category D-3. Other specified secondary infectious diseases
Subclass E. Secondary Cancers
Category E-1. Specified secondary cancers listed in the CDC surveillance definition for AIDS
Category E-2. Other cancers possibly secondary to HIV infection
Subclass F. Other Diseases Possibly Due to HIV Infection

2000 children had symptoms of disease that do not fit the CDC criteria for classification. It is expected that by 1991 more than 3000 children will have the disease (Koop 1987). Epicenters of disease have been identified in the United States; New York City has the highest total number of cases of AIDS, followed by San Francisco and Los Angeles. Of the total number of AIDS cases, 66% are homosexual/bisexual males; 17% have a positive history of IV drug abuse; 8% are homosexual males and have a positive history of drug abuse; 2% have a history of blood transfusions; 1% are hemophiliacs; and 3% of the total cases reported have no risk factor (Quinn 1987).

Since 1981, 47,000 cases of pediatric and adult AIDS have been reported to the World Health Organization with most cases reported from central equatorial Africa. Fifteen to 20% of pediatric patients hospitalized in Zaire have AIDS-related symptoms. Brazil and the Caribbean countries also have large numbers of AIDS patients.

Twenty to 60% of children born to infected mothers will acquire the disease. Half of these congenitally infected infants are black and one-quarter are Hispanic. All will probably die. In many cases the neonate is the index case leading to the diagnosis of the asymptomatic mother. On the other hand, it has been found that pregnancy may accelerate HIV expression in some affected women. Possible risk factors for the transmission of HIV infection from the mother to the infant include viral factors and IV drug abuse. False positive antibody testing of infants is possible due to maternal antibody. Eighty per cent of the infected children have at least one parent with the disease. HIV children are often orphaned or abandoned by both their natural parents and society. Foster parents are needed to help these children lead "normal, dignified" lives.

Presently, approximately 80% of the cases of pediatric AIDS have been the result of transmission by infected or at-risk mothers to the child during birth, 12% by transfusion, 5% have been diagnosed as Factor VIII or IX hemophiliacs, and 3% have no risk factors determined.

Approximately 17% of the children with HIV infection had it transmitted via blood products. The incubation period from exposure to seroconversion is from six weeks to six months. Symptoms appear in the child two to 76 months after incubation (mean of 38 months). The mean age of perinatal AIDS is 2.9 years, predominantly in males. None of the reported cases of HIV infection in hemophiliacs were the result of screened, heat-treated blood. AIDS is rarely seen in pediatric oncology patients or children on dialysis, but there is an increased risk of infection with increased transfusions. Transplacental transmission of HIV has been reported. Infants also have acquired HIV through breast milk infected with HIV from a postnatally infected mother.

Clinical Presentation

A voluminous amount of information has been accumulated concerning the clinical manifestations of pediatric AIDS. The clinical picture of seropositive AIDS in the infant includes: fever, failure to thrive, weight loss, diarrhea, diaphoresis, lymphadenopathy, hepatosplenomegaly, and chronic cough (Koop 1987). A summary of the clinical symptoms of pediatric AIDS is presented in Table 2. Other symptoms from infection with opportunistic pathogens include: persistent oral esophageal candidiasis despite appropriate therapy, lymphoid interstitial pneumonitis, encephalopathy, cardiomyopathy, protonemia, hepatitis, salivary gland enlargement, and recurrent herpetic lesions. Karposi's sarcoma and hairy tongue, although often seen in adult AIDS patients, rarely are seen in infants with AIDS (Quinn 1987).

TABLE 2. Clinical Abnormalities in Infants with Acquired Immunodeficiency Syndrome

Findings reported in > 90%
Poor growth/failure to thrive
Chronic interstitial pneumonitis
Hepatosplenomegaly
Findings reported in 50% to 90%
Diffuse adenopathy
Findings reported in 10% to 50%
Protracted or recurrent diarrhea
Thrombocytopenia
Birth weight < 2500 g
Eczematoid rash
Recurrent otitis media
Developmental failure
Microcephaly
Findings present in < 10%
Kaposi's sarcoma
Chronic parotid swelling

* Table 1 is from the CDC-Classification System for Human Immunodeficiency Virus (HIV) Infection in Children Under 13 Years of Age (draft in press). Tables 2-4 are from Barbour SD: Acquired immunodeficiency syndrome of childhood. Pediatr Clin North AM 34:247-68, 1987.

Another characterisitic of childhood AIDS is a shorter incubation time between HIV acquisition and opportunistic infection. It appears that the infant may develop symptomatic AIDS three or four months from the time of viral infection (Barbour 1987). Chronic lymphocytic interstitial pneumonitis (LIP) diagnosed by histology often is seen in infant AIDS and has been well documented in adults. In a seropositive infant, LIP is a diagnostic feature for AIDS (Parks and Scott 1987). Common bacterial infections occur frequently in childhood AIDS and account for significant morbidity and mortality; these include otitis media, pneumonia, and gram-negative sepsis.

The spectrum of clinical HIV infection is extensive; it ranges from the asymptomatic, seronegative but infected child, through the symptomatic, seronegative child, to the asymptomatic but seropositive child - including children with infectious mononucleosis-like illness reflecting systemic viremia (Barbour 1987). The natural history of the disease is still unclear.

Dr. Robert Marion of the Albert Einstein College of Medicine reported that newborns infected with HIV have identifiable facial features that can be used to help identify disease. The "fetal AIDS syndrome," as the pattern of facial characteristics is called, includes a small head, prominent forehead, flattened nose, and very prominent eyes with a bluish tint in the sclera. It was reported that these traits can be found in up to 75% of the children infected with HIV. It should be noted that the presence of these facial characteristics is still being debated in the pediatric community.

Therapeutic Modalities

Opportunistic pathogens are a frequent finding in children with HIV. Compiled pediatric AIDS cases from the CDC in January, 1986, documented the following data: pneumocystis in 58% of the cases; disseminated cytomegalovirus, 19%; esophageal candida infection, 15%; cryptosporidiosis, 6%; Karposi's sarcoma, 4%; and other opportunistic diseases, 22%. Ordinary infection may be life threatening in these young children. Pneumocystis carinii pneumonia is the most common serious infection in children with AIDS and oral candida or thrush is universal. Tables 3 and 4 summarize the therapeutic modalities currently in use. Vaccines are currently unavailable. The Public Health Service plan for the prevention and control of AIDS includes the following statement: "It is unlikely that a vaccine or therapy to substantially limit transmission will be generally available before 1990."

Oral candida infection is universal in both pediatric and adult AIDS patients, and extension of disease to the esophagus is common. Treatment in children includes oral antifungal agents such as nystatin, ketoconazole, or clotrimazole troches. Intravenous amphotericin has been used when involvement has been extensive. Candida esophagitis may be complicated by concomitant herpes infection (Barbour 1987). Herpes simplex infections are managed effectively with intravenous acyclovir.

TABLE 3. Control and Therapeutic Tactics Against AIDS

A. Prevention
   1. Intensive education programs to reduce risk
      Safe sex
      No shared needles
      Safe blood supply
   2. Deferment of pregnancy among high-risk subjects
   3. HTLV-III/LAV vaccine

B. Treatment of HTLV-III/LAV infection: Chemotherapy against retrovirus

C. Therapy of HTLV-III/LAV infection sequelae
   1. Immunological reconstitution
      Bone marrow transplantation
      Transfer of immune competent cells
   2. Immunologic enhancement
      Cytokines (e.g., interleukin-2 and interferons)
      Immunologic adjuvants
   3. Aggressive treatment of infection and malignancy
   4. New therapy for refractory infections
      Mycobacterium avium-intracellulare
      CMV
      Cryptosporidiosis
      Pneumocystis carinii
   5. Prophylaxis against pneumocystis with trimethoprim sulfamethoxasole
   6. Gammaglobulin infusion
      ? Treatment for lymphocytic interstitial pneumonitis (LIP)
      ? Protection against bacterial and viral sepsis

TABLE 4. Investigational Anti-infection Therapies

Infection	*Therapy*
HTLV-III/LAV	Suramin Ribavirin HPA-23 (antimoniotungstate) Foscarnet Imuthiol 3'-azido-3'-deoxythymidine (BW A509U)
Pneumocystis	DFMO Dapsone
CMV	DHPG (Syntex) *or* BW 759u (Burroughs-Wellcome) (acyclovir analog) Foscarnet
Cryptosporidiosis	Spiramycin

Tables 3 and 4 are from Barbour SD: Acquired immunodeficiency syndrome of childhood. Pediatr Clin North Am 34:247-68, 1987.

Psychosocial Problems and Attitudes

The psychosocial problems encountered by children with AIDS and the frightening picture it elicits sets this disease apart from every other public health disorder (Christ and Wiener 1985). Its onset influences everything important and dear to the patient, his/her family, and friends. Provider institutions, community agencies, and school systems become involved. Ethical issues become a matter of debate and create a schism in the health care field and in the community itself.

The Family

Diagnosis of HIV infection in children creates a state of crisis in the family. It is important to discuss the details of the illness with the family, help them cope with the available resources to treat the illness, and explain the part the family must play in the total care of the patient. The way the crisis is handled can make the family either stronger or weaker (Boland 1987). Response to the initial diagnosis creates shock, disbelief and a sense of denial until the parent can gather the inner strength necessary to cope.

AIDS and its therapies are stressful for the young child and the adolescent. For the parents, not only are the high mortality rate and the young age of the patient stressful, but the debilitating and disfiguring effects compound the stress factors. Patients with a fatal disease often use denial as a defensive mechanism to control their predicament, but AIDS patients find such denial impossible because of constant coverage of the disease in popular broadcast and print media. Denial is intense when the mother belongs to a risk group and is infected herself (Christ and Wiener 1985; Boland et al. 1987). This lack of acceptance can last for months and often will interfere with medical advice, so the child subsequently suffers. Since many parents are carriers, the mother's focus may be on the child's symptoms while not accepting the illness in herself. It becomes quite difficult to maintain a hopeful attitude about the future and the chances for cure.

Young adolescents often are demoralized by newspaper reports. Older children inwardly worry about transmitting the disease to family and friends. Since the general population is somewhat ambivalent about AIDS, patients are denied some of the psychological benefits such as nurturing that other seriously ill patients receive.

It has been reported that at diagnosis, many health care providers fail to recognize the AIDS patients' immediate and long-term need for psychological support (Boland 1987). It is important that psychological intervention by mental health professionals begin immediately. Thus, early intervention will help to resolve how medical treatment will be paid and how the patient will communicate with family and friends about AIDS.

Families adopting children with AIDS need to be aware of the implications of the diagnosis. Especially in those families where parents have succumbed to the disease, foster parents must be found and helped to comply with the demands of the medical care (Boland et al. 1987). The response of the community to the foster family also must be considered. Ostracism and stigmatization may occur when the diagnosis becomes known. Governmental agencies must provide support to the acquired families if placement is to be successful (Ginzburg 1987).

Children in later stages of the disease need help eating, bathing, dressing, and with other basic activities of daily living. These simple tasks we normally take for granted cause a tremendous strain on the patient and the people close to him (Holland and Tross 1985). The overwhelming emotional aspects of these simple tasks often interfere with therapy. Fear often causes the intelligent youngster to place excessive demands on the health care providers and may lead to mistrust.

The feelings experienced by any patient approaching death become aggravated in AIDS children because of the limited knowledge about the etiology, treatment, and transmission of the disease. Therefore, social abandonment results in isolation (Christ and Wiener 1985).

Support systems normally available to medically ill patients which include the family and a network of friends and neighbors are less available to the child with AIDS. Young adult patients with illnesses such as leukemia become intensely reinvolved with their families and rely on them for financial and emotional support. Pediatric cancer patients receive psychological intervention to help them regain a sense of autonomy. Their families try to develop ways of being directly involved with the youngster without undermining his sense of identity. The AIDS child lives in a "family reconstitution" with close relatives (Christ and Wiener 1985). In these children, there is a wide range of mental status from psychological maturity to psychological dysfunction. Children who have psychological dysfunction require psychosocial intervention to permit them to cope with the many ongoing complex problems. Almost constant staff intervention is required to control the periods of frustration and high anxiety secondary to medical status changes.

Because of the stigma of AIDS and their own guilt feelings, parents often fear social rejection and withdraw from society.[1] They feel that the traditional support system of friends, family, and clergy would not understand and that their confidentiality would not be protected. Parents become adept at avoiding questions regarding the child's illness which just prolongs acceptance of the illness by the family.

Family members also have other conflicts about the child with AIDS. When therapy is not successful and death is imminent, families usually wish to be close to the patient, yet fear of contagion arises. At times, the families may request that the AIDS diagnosis be omitted from the death certificate. Each new crisis forces the family to deal with the death of the child. Parents often question the advisability of treating the HIV infection when there is no effective antiviral drug presently available; they don't want the child to suffer needlessly (Christ and Wiener 1985). Each family must be evaluated individually to determine family relationships,

[1] Boland 1987; Christ and Weiner 1985; Holland and Tross 1985.

weaknesses, and strengths in dealing with the HIV infection. Family resources for the duration of the illness must be ascertained (Boland 1987).

Health Care Providers

Health care providers are subject to stress and often find that they themselves require support services because of their involvement with the child with AIDS. The enormous demands on their own energies and the anxiety and fear for themselves and their families of the risk of contracting the disease is a daily stress factor. Staff often become "family" to the child. The Social Work Department at Memorial Sloan-Kettering Cancer Center developed a psychosocial intervention program for patients, friends, relatives, and the center's staff in 1982 (Christ and Wiener 1985; Siegel 1987). The study revealed a major lack of home and supportive care resources and a lack of coordination between acute care facilities and community groups. Techniques for relaxation and behavior showed patients how to cope with anxiety related to medical procedures, pain, and stress. Education and programs for physicians and other professionals helped allay misinformation about the disease.

With knowledge of AIDS and its probable mode of transmission, fears of health care personnel about their own health while treating these patients were reduced. There is, however, burnout among service providers. This is due to the disproportionate increase in the number of AIDS patients and to the limited number of health care providers. Volunteers to help AIDS patients have decreased, and difficult psychosocial problems have not abated.

Community Response

Community response has passed from panic to the establishment of information sharing groups and mobilization of political groups to monitor services as well as advocacy for patients (Christ and Wiener 1985). This has resulted in the resolution of many psychosocial problems. However, the sociocultural burden of a diagnosis of AIDS creates untold problems. The social stigma associated with the contagious aspect causes altered behavior in people including physical and social avoidance. These children become isolated from schools and from other children by frightened parents (Kusknick 1987). The American Academy of Pediatrics has established recommendations for school, day care, and foster care attendance for children and adolescents with HIV infection. Hemophiliacs and recipients of blood transfusions receive sympathetic response due to the perceived random nature of the exposure (Holland and Tross 1985). Even though there is an increase of persons who have acquired HIV via transfusions, the tendency is still to blame the victim. Parents of children with transfusion-related illnesses blame themselves for permitting the transfusion as well as the hospital for not allowing the family to give their own blood.

Confidentiality

Fear of disclosure is pervasive in members of various risk groups (Cassens 1985). This fear has been grudgingly acknowledged by politicians, researchers, and public health officials.

The parents are confronted with how much to explain to the child, fearing that the child will naively explain to his friends that he/she has AIDS. Protecting siblings is impossible and attempts only increase anxiety and stress. Honest answers according to the child's age and ability to understand will help eliminate unforeseen problems (Boland 1987). Counseling referrals are important when the parent cannot handle the situation.

Initiatives in the Pediatric AIDS Area

Surgeon General Koop announced several action plans to be undertaken in response to recommendations developed by conference workshops to remedy some problem areas. He said that because many young AIDS patients are under the guardianship of child welfare agencies, these agencies must be educated about pediatric AIDS including the proper diagnosis of pediatric AIDS cases using the new pediatric AIDS definitions and the risks and benefits of various treatment protocols. He noted that child abuse is emerging as one of the means of transmission.

Koop agreed that pilot studies on the most efficacious methods of newborn detection, including examining core blood, newborn assessment and follow-up, and laboratory distinctions between maternal and infant antibodies, must be undertaken. In addressing the implications of breast milk transmission, especially in developing countries, Koop recommended against discouraging breast feeding. He noted that there is an immediate need for more intensified programs for IV drug abusers.

Koop also noted the need to develop a network of nurturing homes for children with AIDS in order to ensure their access to comprehensive services. He added that he will bring together representatives from public and private sector organizations to discuss possible partnerships which will guarantee the availability of and access to these services.

Koop cited the need for better minority educational efforts and indicated that a project for reaching minority leaders is already underway (Brown 1987).

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Computers claim new victims

Man (and woman) keeps inventing new pathological conditions. Physicians at the Children's Hospital of Philadelphia have the honor of being the first to report on, and coin the phrase "joystick digit", a variant on the established diagnosis, trigger finger. Obviously, this newest pathology is a child of the computer age. Osterman et al. describe two cases of young males who complained of having a single finger lock in flexion, with flexor tenosynovitis. Both patients admitted playing video games using joysticks for prolonged periods. Unlike the longer airplane joystick, the smaller computer joystick presses directly against the flexor sheath, resulting in the condition described, following prolonged use. Treatment includes anti-inflammatory medication and limited and/or modified joystick use.

Osterman AL et al.: Joystick digit
JAMA 257:782, 1987