Pediatrics Paper: And Then There Was One
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**Introduction: HIV/AIDS**

Human Immunodeficiency Virus and Acquired Immune Deficiency Syndrome (HIV/AIDS) was first recognized in 1981 and is believed to have originated from human contact with the blood of hunted chimpanzees infected with the simian immunodeficiency virus (Centers for Disease Control & Prevention, 2012). Found in the blood, semen, vaginal fluid, and breast milk of infected individuals, HIV has various modes of transmission. Unprotected sex, sharing of hypodermic needles, and mother-to-fetus or mother-to-infant transmission are examples of how the virus can be passed from one individual to another (NIAID, 2009).

HIV/AIDS is a huge public health issue affecting adults and children all over the world, however it is especially of concern among the world’s poor and in developing countries where access to health education and treatment is generally more limited (Gari, Blaya, Hall-Clifford, & Frasier, 2008). Poverty is both a risk factor for the contraction of HIV as well as a barrier to its treatment. Because children are at an increased risk for poverty due to their dependency on adults, they are more susceptible to HIV and its effects (Marais et al., 2008). Perinatal transmission is the most common mode of HIV infection in children (Centers for Disease Control and Prevention, 2013). According to the World Health Association, in 2011 there were 3.3 million children under the age of 15 living with HIV, 330,000 newly infected children, and 230,000 who died from AIDS. Of this 3.3 million, over 2.2 million lived in Sub Saharan Africa (WHO, 2011).
Pathophysiology of HIV/AIDS

It is believed that mother-to-child transmission of HIV occurs in the late stages of pregnancy. While the exact means of transmission is not clearly understood, two hypotheses exist. Exposure to the fetus at the time maternal blood enters into fetal circulation is one theory, and exposure via vaginal mucosa at childbirth is another. Breastfeeding is a means of transmission from mother to infant (NIAID, 2008).

There are three main phases of HIV: primary HIV, latency, and AIDS. These phases are based on the body’s CD4+ T cell count and the development of opportunistic infections, and can occur over the course of 8-12 years (Porth, 2007). Once transmission occurs, HIV attacks the body’s CD4+ T cells and begins to quickly replicate and implant itself in the body’s organs and lymph system. CD4+ T cells are important in maintaining the body’s immune function and fighting off disease. As this happens, CD4+ T levels drop and the viral load of HIV increases (NIAID, 2011). During this primary phase, the infected individual may experience fever and flu-like symptoms. If started on treatment at this point in the infection, the number of long-living HIV cells (CD4+ memory cells) can potentially be reduced. In the latent phase, which can last approximately 10 years, the infected individual is asymptomatic however may experience lymphadenopathy as CD4+ T cell levels drop (Porth, 2007). Eventually, viral load will increase to levels such that the immune system becomes worn out and can no longer defend the body from outside infection. A final diagnosis of AIDS is made when CD4+ T level are less than 200/cubic millimeters of blood, and one or more opportunistic infections are present. The HIV infected individual can live for several years before becoming symptomatic and the diagnosis of AIDS is made (NIAID, 2011).
While about 80% of perinatally infected children generally live into school age and adolescence before acquiring AIDS, about 20% will die before age 4 after becoming seriously ill within their first year of life. The reason for this variability in the severity of the illness is believed to be related to maternal Vitamin A and CD4+ T-cell levels during pregnancy and CD4+ T-cell levels present in the infant at birth (NIAID, 2008).

**Differential diagnosis**

In the movie, “And Then There was One” the unfolding of the diagnosis began when the child Miranda came down with what appeared to be cold and fever. When Miranda’s fever got to 101 F, Roxy placed a call to the MD who told her it was probably respiratory flu and that “all the kids have it”. After a few days, when the fever had not resolved, Roxy and Vinnie brought Miranda into the doctors office. While in the waiting room the parents were discussing how Miranda had gone from screaming for 12 straight hours to not crying at all, that is when they noticed both her fingers and mouth were blue.

The doctors admitted Miranda to ICU and diagnosed with viral pneumonia and promptly started treatment. After twenty-four hours with no response they decided they needed to change antibiotics and “change to a respirator”. She began to improve slightly over the following days and then suddenly both lungs collapsed, Miranda was then sedated and intubated. At the recommendation of the nurse, Miranda was transferred to another hospital because they had access to an ECMO, an extracorporeal membrane oxygenation machine. This would offer her heart and lungs a chance to rest. After a short time on the machine the pediatrician made a diagnosis of PNA Carinii leading to the diagnosis of AIDS.

As a practitioner diagnosing baby Miranda who walked into our office, we would consider working up the child to find the etiology of her respiratory condition. First we would want to do a
thorough history about Miranda with Vinnie and Roxy Ventola. We would want to know questions such as: When did this infection start? Has anyone else around her been recently sick? Has it become better or worse over the previous days? Does she feel better or worse? Is this the first infection or has she had multiple respiratory infections in the past? As for looking at the physical exam: How did the child present, was it a fast onset with rapid fever, labored breathing, and cough? How do the lungs sound? Is there any accessory muscle use, flaring of the nostrils, or bluish nails or lips? Do the vital signs show any indication of current infection such as increased pulse rate, increased respiration, decreased blood pressure, or fevers

Once the H&P exam is complete, we then would begin to work up baby Miranda. We would want to get a CBC to assess WBC’s, blood cultures, and chest x-ray. Based on what we know, we would begin to treat Miranda with supplemental oxygen and antibiotics as a precaution until we determined the source of the infection. After Miranda’s lungs collapsed we would know it was a complicated case and would bringing in specialized physicians to assist with our patient’s needs. Further testing to determine the source of the infection would be necessary to ultimately lead to our diagnosis of AIDS. Although far from thinking AIDS at the time her lungs collapsed, determining the source of the infection to be fungal PNA (pneumocystis PNA) would be the key to leading the diagnosis. A fungal PNA only affects individuals with a poor immune system, we would therefore want to start searching for a disease that would cause an 8-12 month old to be immunocompromised. As pertaining to the movie and diagnosis for children, currently HIV testing can be done between 3-15 months of age, however it is hard because babies carry their mother’s antibodies for several months. A polymerase chain reaction (PCR) test can be done soon after birth that can detect genetic HIV material. Testing the parents rather
than the baby should be done to help make a definitive diagnosis (Avert, 2013). As with all diagnosis, this would be a process of trial and error and ruling out other causes before ultimately reaching a final of AIDS.

In the case of Miranda, since the Acquired Immune Deficiency Syndrome was new and prominent during the 1980’s, a diagnosis of AIDS would have been more on the forefront of the MD’s minds. Although severe respiratory complications such as pneumonia are a common presentation for AIDS, in 2013 this would not be a first choice on the list of differential diagnosis in the beginning. This disease can be difficult to diagnosis especially before opportunistic infections begin to present. As stated above, AIDS itself is not the cause of immune changes; they are caused by opportunistic infections that appear when the CD4 count drops below 200 cubic millimeters of blood and the body no longer can defend itself.

The problem with clinical presentation vs. actual differential diagnosis is that an opportunistic infection can lead to a differential diagnosis of all kinds of simple diseases. Opportunistic infections can affect most body systems and appear to be a simple infection. Most likely a simple list of common differential diagnosis will be first presented before moving on to the more severe and intricate diagnosis presented in the Ferris book (2013).

According to Ferris (2013) the most common differential diagnosis for AIDS is: TB, Neoplasms, Disseminated fungal infections, malabsorption syndrome, depression, and other disorders with dementia or changes within the myelination causing encephalopathy, myelopathy, or neuropathy.

As research suggests in 2013, 21% of Americans living with HIV do not realize they are infected with the disease (NIH, 2013). Unless diseases CD4 count has reached levels where the body
can no longer defend itself, it would be hard to make this diagnosis. However, as practitioners in
today’s society looking at high risk lifestyle choices (multiple partners, sharing of needles) and age (with peak incidence being between ages 20-49) may help lead you to test for HIV before symptoms appear in an adult.

**Interventions**

**Pharmacological**

Although there still is no cure for AIDS at this time, there are treatments that keep symptoms at bay and improve and lengthen life expectancy. In the movie, the Ventola family were plagued by AIDS when very little treatment options were available in the late 1980’s. From what was communicated in the movie, the AIDS specialist prescribed medications for all three members of the Venola family. Vinnie, Roxy, and Miranda were treated with AZT therapy and were given a family informational packet about AIDS.

Although treatment options have opened tremendously since the time of the Ventola family, we are still running into treatment challenges within the pediatric population. Through research, it has been proven that treatment of infants with immediate antiretroviral treatment (ART) decreases mortality and morbidity and may possibly improve neurodevelopmental outcomes. However, it is still unclear if all children under the age of two should be started on ART (Penazzato, 2012). Optimizing dosing of ARV (antiretroviral) is challenging in children. As of 2012 there is still not a good understanding of correct pediatric dosing and toxicity. There have not been enough pharmacokinetics studies completed in children to make a decision on what is safe. According to WHO and UNAID’s guidelines published in August of 2012, HIV and AIDS medication treatment is different for the pediatric population, especially
under the age of three. The first line drug treatment is uptake of LPV/r (Lopinavir). This current regimen is in liquid form and has a foul taste and a high alcohol concentration (up to 42%) (WHO, 2013). It is also challenging to distribute in third world countries because it needs to be refrigerated.

Another option is EFV (Efavirenz) recommended as an NNRTI (non nucleoside reverse transcriptase inhibitors). It is a first line therapy due to its antiviral activity, long half life and ability to be used in conjunction with TB therapy (WHO, 2013). However, the medication is variable and difficult to dose therapeutically in children. There is also fear of resistance due to higher clearance rates in younger children. Again, dosing for children under 3 has not yet been established.

WHO believes there needs to be more protease inhibitor (PI) treatment options for children. They believe ATV/r (Atanavir) could be a good first line treatment in infants due to its lower costs and once a day dosing. Unfortunately, at this time ATV/r and another PI DRV/r (Darunavir) are only approved for children over the age of 6 in the United States.

Within this evaluation and various guidelines, the recommendation stated by PAWG (paediatric antiretroviral working group) emphasizes the importance of pharmacokinetic studies in children and development of more pediatric options for children under the age of 3 (2013).

**Non-pharmacological**

Non-pharmacological interventions and education are an important component for the care of the individual with HIV/AIDS. Achieving a full understanding of what the disease is and how it affects the body will give a greater understanding of how better to improve and protect overall health.

Maintaining good caloric intake is important as the individual’s body begins to waste and become more cachectic. Giving specific guidelines can help the patient stay at an optimal weight which
will in turn help maintain their health. Another way to protect is to receive all the appropriate vaccinations (not live attenuated) to protect from preventable infections. Patients should also be encouraged to continue to monitor their viral load and CD4 count so as to monitor for disease progression. When the CD4 levels reach below 200 -when AIDS is diagnosed- there is a risk for opportunistic infection. At this time, making sure the patient knows to protect themselves from exposure to potential deadly diseases by minimizing public exposure, and avoiding undercooked foods is important (Avert, 2013).

**Palliative**

Although treatment options have come a tremendous distance since the beginning of the AIDS epidemic in the 1980’s and individuals afflicted with HIV/AIDS are living longer, more productive lives than they were able to years before, there is still a need for palliative services. As the disease progresses causing CD4 counts to drop, viral loads to increase, and individuals to become more susceptible to opportunistic infections, the need for palliative care becomes more beneficial. Palliative care is an interdisciplinary approach used to encompass the whole patient while alleviating uncomfortable symptoms and achieving a higher quality life for the patient (UCSF, 2013). The team, consisting of an MD, nurse, nursing assistant, social worker, chaplain, and volunteer, can aid the patient and family through the difficult end times dealing with physical, psychosocial, and spiritual needs.

In the case of the Ventola family in “And Then There Was One”, a palliative care team could have been of great assistance to Roxy. Not only did she have to work, but she also had to care for her ailing husband Vinnie and daughter Miranda both stricken with the AIDS virus. The nurses could have come to the home one to three times a week to draw the families CD4 and viral counts, manage any
symptoms with medications, and observe everyone's general health. A nursing assistant could have assisted Vinnie and Miranda with their ADL care and general housekeeping items. The social worker and chaplain could have assisted Roxy with her emotional and spiritual state, allowing an outlet to talk and ask questions. There could even have been a volunteer to give Roxy a few hours break to allow her to work or give her the option to leave the home for errands or a much needed break. Although this didn’t appear to be an option for the Ventola family at the time, palliative care would have been highly beneficial to them.

Human Immunodeficiency Virus and Acquired Immune Deficiency Syndrome have devastating complications in both adults and children. The implications of these diagnoses were highlighted in the film “And Then There Was One”. The hardships of Miranda were especially apparent, being a young girl coming to grips with her own diagnosis of HIV/AIDS, as well as that of her family. HIV and AIDS have a complex pathophysiology that is still being researched. Pharmaceutical drugs for the treatment of HIV/AIDS have developed considerably since the 1980’s. However, medications for the pediatric population have lacked research and haven’t made much progress to date. Nonpharmaceutical interventions are important to manage nutrition, weight, and educational needs. As a nurse practitioner evaluating a child with symptoms, a careful history must be taken to identify the risk factors of the parents and to establish a possible differential diagnosis. HIV/AIDS is a devastating diagnosis that requires a complex plan of care and management of the patient and the whole family.

References


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