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Thalassemia and Asian Americans:
Living and Coping with Uncertainty

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Abstract

Thalassemia is a potentially life-threatening genetic blood disease for which Asians in California are at highest risk, compared to other population groups. Mandatory screening at birth is how most cases are discovered. This paper focuses on chronic forms of thalassemia and what it means for patients and their families to live with the illness. The goal is to increase public awareness about thalassemia and to stimulate discussion about social interventions that might enable individuals to lead healthier lives.

Introduction

Thalassemia is a hereditary anemia that is among the most common of genetic disorders (Rund and Rachmileqitz 2005). Historically the disease has been associated primarily with Southern Europeans and the geographical area implied in the name of the disease—"Mediterranean anemia" or "anemia by the sea" (*Thalassa anaemia*) (Heer et al. 1998). Thalassemia is now known to be prevalent among those of Mediterranean, African, Middle Eastern, and Asian/Southeast Asian descent. The mutations are believed to be environmental adaptations protecting against malaria as well as certain non-malarial diseases (Allen et al. 1997).

In California Asians are the population at highest risk for thalassemia, the combined effect of high rates of Asian immigration/migration to the state, high fertility and the tendency towards marrying within these same communities, and the introduction in 1990 of mandatory newborn screening for hemoglobinopathies (Lorey 2000; Lorey and Cunningham 1998; Heer et al. 1998; Nidorf and Ngo 1993; Anderson and Ranney 1990). In its more serious and chronic forms (beta thalassemia major and Hemoglobin E), the disease is one where noncompliance with medical protocol is a major cause of death among young adults (Modell and Kuliev

1993). With compliance, the annual costs of lifetime treatment are significant. In 1995, the estimated cost of care for a transfusion-dependent child was $69,500 per year, resulting in a total lifetime cost of $2,432,500. A decade later, these annual costs have increased more than five-fold. An adult with beta thalassemia in 2005 would have medical care costs of $366,809 per year.

Prompt and early care can optimize life expectancy. This paper provides a window into the social and cultural factors that affect access, use, and quality of health care services, with the focus on how chronic forms of thalassemia affect quality of life of patients.

"What is Thalassemia?"

As a blood disorder, thalassemia constitutes a group of genetic mutations that impedes the oxygen transport component of red blood cells. Infants usually appear healthy at birth but begin to show signs of anemia between six months and two years of age. Left untreated, most patients die by age five (Modell and Kuliev 1993). Treatment involves red blood cell transfusions every three weeks, which eventually lead to iron buildup in different organs. Iron overload is the source of most morbidity (e.g., diabetes, liver problems) and mortality (e.g., heart failure being the primary cause of death). By removing the excess iron, iron-chelation therapy can double life expectancy (Rund and Rachmilewitz 2005).

For the most part, a well-cared for child with thalassemia does not look different from any other child and can function normally. Most patients are compliant with blood transfusion because without it, they feel tired and run down. Compliance with iron chelation therapy, on the other hand, is much more problematic because the regimen is itself onerous (involving subcutaneous infusions of the drug for several hours a day, eight to fifteen hours a day, five to seven days a week). It can take years before patients suffer the effects of noncompliance with treatment procedures.

The only available cure for thalassemia is bone marrow stem-cell transplantation, ideally with a fully compatible sibling match (Rund and Rachmilewitz 2005; AAPCHO 1999). Such a procedure is itself risky with potentially serious complications (e.g., death, graft failure or rejection). In 2005, the estimated cost for bone marrow transplant at the Children's Hospital and Research Center in Oakland was $500,000.[1] Gene therapy and pharmacological cure are still very much in the distant future.

Data and Analysis

Learning how genetic information is incorporated into the lives of patients and their families was a major objective of this study. Although life expectancy for thalassemia patients in the U.S. is greater than that in Third World countries where blood transfusions themselves can be unaffordable, less than full patient or family commitment to the prescribed medical care can still prematurely shorten lives or adversely affect the quality of life. In the early 1990s, the thalassemia program at Children's Hospital Research Center in Oakland (CHRCO) was in the early stages of development. The research project itself evolved out of a larger study at CHRCO that focused on the social and cultural barriers to genetic testing and screening on populations at risk for sickle-cell anemia and cystic fibrosis, respectively, African Americans and European Americans. Clinicians working with thalassemia patients asked to be included in the interview project, motivated largely by the hope that the study's findings would enable them to improve the level of patient or family participation in the program.

Specifically, the research team sought to learn about the following six topics: history of personal experience with the genetic disorder; perspectives on the disease, including beliefs about its meaning, prevention, and treatment; knowledge about the availability of genetic screening and attitudes towards carrier testing and prenatal diagnosis; family communication; communication with friends and acquaintances regarding the disease; and health care concerns, including those related to insurance and coverage.

These topics were explored through interviews conducted within the hospital setting when patients were admitted for blood transfusions. Since the thalassemia project was unfunded, there was no money for transcription, let alone incentives that could be offered to remunerate families or patients for their participation. The goal of moving beyond this initial point of hospital contact into the extended families was rarely possible given time pressures on parents and the need to care for children. In short, the data were derived mainly from formal or informal interviews in the clinic, as well as field observations here and, to a very limited extent, outside the hospital. There were also methodological challenges, which I will come to shortly. But despite these circumstances, the project team conducted fourteen formal interviews with patients or their

parents, the bulk of which took place between 1993 and 1995. Field notes were kept on informal clinic conversations, while the formal interviews were mostly (but not always) tape-recorded and lasted on average about an hour, with some significantly shorter. In total, the team observed thirty-five "cases," which were officially part of the thalassemia project. The majority were parents, sixteen mothers and ten fathers, and the remainder patients. In 2005, I revisited the project, conducted select interviews with the research/medical team, and drafted this analysis.

Although the interview project also included three Iranians, an Afghani, an Italian, and a Greek American, the thalassemia "cases" were largely Asian, and hence the findings relate to this population. The majority of the Asians were either first or second-generation immigrants, many of whom were Southeast Asian but diverse in ethnic background (Chinese, Vietnamese, Laotian, and Cambodian) and religious orientation (Christian, Catholic, Buddhist, "no religion"). In terms of socioeconomic status, the majority of Asian patients had family incomes below $50,000. Only two interviewees had family incomes over $50,000 while family incomes for the rest fell below that: $30,000 to $49,999 for those with some college education, and below $10,000 for those who were either unemployed or else marginally employed.

The lack of funding prevented training available interpreters for this project. Furthermore, since the research team was made up of individuals recruited originally for the larger study, there had been little planning as to how to bridge issues like "cultural distance." Although not part of the original research team, I was later invited to join the project to assist with rescuing a data collection process fraught with certain conflict between members of the research team. Conflict arose over the framework in which to interpret the experiences of patients and their families, specifically over whether they were better understood in terms of specific struggles with the *medical* regimen, or in terms of the social and cultural *meanings* that framed this medical experience. Although both issues are substantively important *findings*, they became fundamentally opposing *orientations* as to what the project was "about." This conflict had direct implications for how systematically information would be collected as well as how descriptively rich or "thick" that information would be. Despite the challenge of analyzing data under these less than ideal circumstances, I produced a draft that

bridged some of these differences and ultimately became part of the project's Final Report (Duster and Beeson 1997).

The present paper develops that framework, following the model offered by Rose Weitz's study of persons with AIDS. The model focuses on coping under the stresses of *uncertainty*, an issue which medical sociologists have deemed an important issue for chronically ill or terminally ill patients. Weitz' model identified two competing orientations towards uncertainty: (1) a "vigilant" coping style that entailed actively monitoring one's condition, e.g., seeking information to make better health decisions, exploring alternative treatments, other expert opinions, and, any promising proactive avenues that would restore a sense of individual control; (2) "avoidance" strategies that emphasized ignoring or minimizing exposure to negative or unpleasant information, e.g., preferring the uncertainty of not knowing to the certainty of a positive diagnosis, avoiding contact with physicians, downplaying symptoms or attributing symptoms to less serious conditions, and listening only to optimistic accounts (Weitz 1994).

In the thalassemia study, these patterns first appeared around the diagnosis ("why me?" or "why my son or daughter?"), and subsequently around treatment care and communicating genetic information to others. This paper describes how parents responded differently to the social and medical uncertainty created by the diagnosis, as well the context for their responses.

Thalassemia patients differed from AIDS patients in important ways. Weitz theorized that AIDS patients, i.e., gay males, would suffer from uncertainty to a far great extent than other patients for several reasons: (1) they could reasonably anticipate their chances of being at risk, thereby experiencing significant emotional turmoil prior to diagnosis, (2) they would likely experience guilt for behaviors contributing to their illness, (3) face difficulties in obtaining accurate diagnosis, (4) experience greater uncertainty in how illness would affect their lives, and (5) insofar as AIDS was a new disease at the time, lack answers to their questions about treatment and prognosis (Weitz 1994).

While the state of medical knowledge regarding AIDS is now more advanced than it was a decade ago, the areas of uncertainty outlined above provide a useful framework for analyzing the experiences of families with thalassemia. (1) Unlike AIDS patients, families with thalassemia typically lacked any anticipatory fear re-

garding their own risk level. Quite the opposite, they were shocked to learn about a "genetic" disease. If previous family members had died from the disease, it had never been recorded as such. (2) As with AIDS patients, guilt *was* a prominent theme, but unlike the situation of AIDS patients, the medical explanation had the potential of alleviating some of this guilt (Parsons 1951). (3) Genetic diagnosis offered greater "certainty" (at least as far as diagnosis) than did medical knowledge in the case of AIDS a decade ago, but (4) there remains a tremendous amount of uncertainty as to how to normalize a life style that now included living with thalassemia. (5) Finally, within the realm of public awareness and even among medical doctors, thalassemia is still a "new" disease about which little is known, and for which medical treatment continues to be largely "maintenance."

Findings

"Avoidance" as a coping style

The initial reaction to a diagnosis of thalassemia is typically shock and resistance to the idea that "genetic" disease runs in the family. A 42-year-old mother encountered not only disbelief and denial from her in-laws but also blame. As in the following quote, patients, families, and staff alike commonly use the more colloquial term "thal."

> Mrs. X's in-laws. . .did not believe this explanation. They thought that it was all Mrs. X's fault that she and her husband had a sick child. Her mother-in-law in particular blamed her alone for the problem saying that her child was sick because she was so skinny. Mrs. X also said that her husband's brothers and sisters did not believe that it was a disease that ran in their family until three years later when her husband's brother had a child with Thal. At this point her husband's younger sister went to get tested before she got married.
>
> Mrs. X ran into similar problems with her own family. . . .They didn't believe that the disease ran in the family or that they were at risk for it and they did not get tested initially. Now, however, they have gotten tested.
>
> (Arona Ragins, field notes, March 31, 1995)

Efforts to make sense of the diagnosis ("Why my children?") occur at both a medical and metaphysical level. Although the affected families sometimes sought alternative diagnoses, the single

major cultural belief that competed with medical understandings was a belief in karma. The belief, moreover, encouraged a strategy of doing nothing about the illness but rather accepting it as part of one's fate.

> A child born with thalassemia major is a Karma child, working out a problem of the ancestors. If the child dies early, then the problem may be considered resolved. . . .Suffering is a natural part of existence. . . . One must be stoic and accept one's fate. . .To intercede is to create disharmony in the universe and anger God. (Nidorf and Ngo 1993: 399).

Parents experienced great remorse, believing they were paying the penalty for actions in a previous lifetime. Thus, a Chinese father suffered much guilt over his own possibly contributing role. The field researcher said about this thirty-year-old waiter: "He used to blame himself for the fact that (the child) has thalassemia major. He sometimes thinks that he did something bad in another life and that now he is paying for it by having a child with problems." Similarly, a Chinese mother, a twenty-eight-year-old phone receptionist at a daycare center, initially concealed her child's illness for fear of being ostracized.

> . . .that there was something bad in their family that they had done to deserve this. There is a cultural belief/superstition that imperfect children were seen as punishment for bad associations in the past such as among the ancestors through the family line. (Woo, field notes, September 13, 1993)

There is also a gender-based source of maternal guilt, as mothers regretted following their doctor's advice to get prenatal testing. One mother even believed there were early warning signs during her own pregnancy to which she ought to have paid greater attention. Mr. A, the husband, thus explained:

> . . .for the longest time for about three to four years she cry, you know, when she sit alone. . . I caught her crying. And then we talked about it. . . (even though) she has enough understanding of thalassemia as a disease or disorder, does that mean any easier for her to accept the disorder? . . .prior to, to each of the child, to, to my son and my daughter (who has thalassemia), my wife had miscarriages. . . .she was very healthy female, so the fact that she had miscarriages as often as she did. . .she still continues to feel kind of guilty about that. . .
>
> (Chinese male, 43 years old, engineer)

While medical diagnosis can mitigate a good deal of guilt or shame, misconceptions about disease can exacerbate the problem. The disease may be understood as having to do technically with "something in the blood," but blood transfusions raised fear of contracting AIDS. One family adjusted to this perceived risk by separating the sick child's food and utensils and by keeping the child at home. Social isolation, however, could be more emotionally wrenching than the disease itself. When his daughter began transfusions, one father described this as "the most difficult phase in my life." They became pariahs in the eyes of "openly hostile" relatives fearful of contracting disease through social contact. These relatives are now quite accepting of the daughter and her condition, likening thalassemia to other chronic conditions (e.g. arthritis) which are not "contagious," and even seeing it as less debilitating than other health problems (e.g. mental retardation). Fear abated, however, only when accurate, public health information about AIDS became more widespread.

Resistance to genetic diagnosis aside, three major factors worked against compliance with the treatment regimen: the arduous nature of the treatment, an impersonal hospital bureaucracy, and otherwise normal patient functioning. Given this situation, it is perhaps amazing when patients and their family members do cooperate. Genetic diagnosis alone is of limited value in promoting compliance. An otherwise healthy appearing child made it hard to convince parents to seek treatment. This resistance to seeking help when there are no outward, visible symptoms of a problem is a dominant cultural orientation among Asians, shaping attitudes towards even simple medical procedures, including newborn screening for thalassemia (Nidorf and Ngo 1993). Many patients do not appear to need blood transfusions (Eleftheriou 2003). Thus, in the early stages when parents were learning about thalassemia, few could appreciate the benefits of blood transfusions. The provider interested in promoting long-term patient self-care cannot simply impose clinical necessity but rather may have to forgo treatment until parents or caregivers acquiesce. Dr. Elliot Vichinsky, Director of Hematology/Oncology at Children's Hospital and Director of the Thalassemia Clinical Regional Network, respected the family's right to decide but was equally diligent about educating caregivers on the possible consequences of refusing treatment. Over time, parents learned to associate the fact that an infant who had become

visibly ill was in need of regular transfusion. In this way, respecting a patient's or family's choice, however risky, but continuing to work closely with them promotes not only better understanding of the disease but greater trust in treatment programs essential to long-term care (Lerner 2006).

Another treatment option, self-administering the drug Desferal, involves daily needle injections and wearing a cumbersome unit around the waist for about twelve hours every day. While the beneficial effects of a blood transfusion are immediate and visible, the ill effects from not doing Desferal may not be experienced for years. Outreach worker Laurice Levine is also a thalassemia patient who regards herself as among the more "vigilant" types and thus lives a very high quality life. At the same time, she understands how patients can be remiss or negligent, explaining:

> You feel awful if you don't get blood – tired, exhausted, out of breath, weak, dizzy. When you use Desferal, you feel side effects like a bruise, or a bump, or a lump, but when you do not do Desferal, there are no immediate side effects. If you didn't do Desferal for a week, you wouldn't feel any worse than if you did do it. . .It can be years before you experience the side effects of not doing Desferal. So you don't get any instant return. . .there's no kind of payback. (Levine 2005)

Many young Asian adults with thalassemia come from struggling homes and poor communities that would be considered disorganized or dysfunctional by middle-class standards, lacking the support structures that would provide a child with stability, a sense of purpose and discipline. As with other chronic sufferers of seriously debilitating illness, the illness experience is but one feature of their lives (Conrad 1994). Levine thus explains the challenges.

> Some patients have poor family lives and few resources and some have very little support at home. There are so many patients who have troubled lives besides thalassemia and I think that's a huge factor in compliance. Mere survival is a challenge, let alone having to commit to an intense medical regimen.
>
> I think that support wouldn't even mean, yelling at the child about Desferal. It would mean an organized household, moral support, good role modeling about work ethics, about living life to the fullest, getting along with your neighbors, cleaning up after yourself. (DW: Nothing about the disease itself?) No

> because that just turns patients off. The more you tell them to do it, the less they want to do it. I know people who have cut the needle off and taped the pump to their stomach and they look like they're running the Desferal but they're not running the Desferal. So the needle didn't go into the abdomen, got snipped off, and they taped it there. So I think it's just having a really good household, and a good family life, and a good neighborhood (Levine 2005)

In short, those families where noncompliance is the norm are likely to be overwhelmed by numerous life events. Thus, a Cambodian mother, distressed over her son's gang affiliations, delinquency, school problems, and running away from home was described by a field researcher as "not sure whether his problems are because of his *thal* or because of his being a teenager." In the case of a fifteen-year-old Cambodian female, who missed numerous transfusion appointments, the medical staff considered more family conferences, counseling, and possibly contacting Child Protective Services. The suggestion that these issues be put in writing in both Cambodian and English indicates a lack of basic mutual understanding.

If patients' family lives seemed disorganized, the hospital bureaucracy appeared impersonal, insensitive, and unresponsive to their daily life circumstances. Patients and family members often had to take time out of busy schedules, in some cases traveling an hour's distance, for what seemed to them to be trivial or inconsequential exchanges, better accomplished by phone or some other efficient means. Those who were able to adjust did so only because of their own coordinated efforts to accommodate medical appointments.[2] Once within the clinical setting, long waits might still be in order. One research assistant accompanying a thirteen-year-old undergoing medical tests was struck by the inefficient way medical data were gathered, repeatedly bombarded as the patient was by the same questions.

> I was amazed at how many questions "H" (the patient) would be asked during the next hours—literally dozens of health-related questions, instructions, and clarifications asked by every practitioner he came across—nurses, doctors, techs, etc. Many of these questions were repeated by different clinicians in different settings. (Jason McNichol, Field Notes, June 23, 1994)

In general, delivery of care was perceived to be fragmented and incongruent with other aspects of their lives. Thus, although severe anemia might be one explanation for the "listless" or relatively unresponsive style of patients, it was also true that the hospital bureaucracy took its own toll. In this environment, patients were estranged and alienated, so it is not surprising that members of the research team described them as "sullen," "quiet," "aloof," "distracted," "suspicious," "uneasy" "bored," or "withdrawn."

Language barriers alone can discourage individuals from seeking professional help. In one large extended family, the divorced Laotian mother of a 13-year-old with thalassemia communicated little about the genetic disease to her mother or twenty brothers and sisters, despite the fact that they were emotionally close and saw each other on a regular basis. No one, moreover, had sought testing for the genetic trait, even though the mother herself had been a health practitioner in Laos and two of her brothers were doctors. According to her companion, a 47 year-old white male who served as the primary caretaker, the failure to follow through with genetic testing was due to language barriers and lack of education. Where parents were more highly educated, vigilance was more the norm.

"Vigilant" styles of coping

Vigilance as a coping style reflects a proactive stance towards the illness, one facilitated by higher education. Better medical understandings of the illness can facilitate destigmatization, yet fatalistic attitudes and fear of social rejection can still discourage one from seeking tests or treatment. Mr. T., a 45-year-old native of Hong Kong, for example, had learned about genetic inheritance patterns in high school, and all Hong Kong citizens were encouraged to get genetic testing before marriage. Yet fearful of being rejected by his future wife, Mr. T. did not go in for testing, with the result that he is now the father of two boys with thalassemia. Moreover, he resisted treatment given his fear that this would "extend his sorrow."

Mr. T's attitude shift from avoidance to vigilance once it became clear that transfusions were helpful.

> Initially Mr. T was reluctant to have his son transfused because he held the belief of traditional Chinese philosophy that things were the way they were meant to be. He felt that treat-

> ment would only extend his sorrow. His son, however, began to receive transfusions and after that Mr. X changed his mind about the treatment doing more harm than good because it was obvious that the child felt much better when he was being transfused. (Arona Ragins, field notes, February 2, 1995)

The positive medical experience let to his actively advocating genetic testing for other ailments (e.g., Down's syndrome or other developmental disabilities), to investigating the family history, asking even his parents to get tested just so that he could "find the truth." (They refused, offering the practical reasons many others also gave, that they were not having children and saw no need.) In short, Mr. T is now "not ashamed to talk about *thal* at all," explaining that "it's no one's fault."

A quintessential example of vigilance was Mr. A, whose wife was mentioned in the previous section as weighed down by tremendous guilt.

> His principle strategy of dealing with her disease was to gather extensive information about treatment procedures – reading technical articles, discussing the matter with doctors, and becoming, in the end, an expert himself. He consulted with all of the top practitioners about the disease traveling to Seattle, New York, and even Italy to do so. (Arona Ragins, field notes, August 26, 1993)

Thus, while most patients adopted an avoidance strategy in response to the arduous nature of the treatment, the same situation propelled Mr. A to aggressively search for some permanent cure. Prevention was an extension of this proactive stance. He thus explained the paramount importance of informing younger family members so that they would have greater options.

> . . .I have no hesitancy talking to them. . .my brothers are still all very young. . .an age at which they will be getting married, having kids. . . . If they don't know about it, I want to talk to them. Put it this way. If we were smart enough to do amniocentesis in the beginning, (we'd) probably terminate the pregnancy.
>
> (Chinese male, 43 years old, college-educated)

Tracing the history of the disease through the family tree through contacting elder family members was also pursued, though eventually dropped because there were no direct implications for immediate health care or preserving some semblance of normalcy in a child's life. Mr. A thus actively consulted his daughter about

how she would like to relate to individuals outside the family, offering up the example of how they addressed certain issues related to physical education (P.E.) class.

> We work with (the child). . .We ask her whether she wants to tell her friends that she has thalassemia. We talk to her together with her pediatrician (about) whether we ought to inform the school nurse that she has thalassemia in case an emergency comes up. . .And we then coach her (about) how she should say to people, and, what thing(s) not to do.
>
> . . .she has marks on her tummy from the Desferal so she, she doesn't wear two-piece swim suits. She wears one single piece. . .she get tired easily, so (we) tell her, "tell your P.E. teacher you only run so fast. . .that you are anemic," which is quite normal. . .I say, when you are ready to tell your friends, ah, you come and you talk to us about it and we'll deal with it in that manner. It's her choice.

Still another vigilant father, Mr. C, communicated a great deal about his son's illness with others, including nonfamily members, because this was critical to mobilizing practical support.

> Unlike some other Asian families in our study the (family members here) communicate freely about *thal* with non-family members. Mr. "C" said that "Y's' teachers know about his *thal* as do some of "Y's" friends. One of "Y's" friend's mother, who is a nurse, even asked Mr. "C" to teach her how to administer a Desferal pump. "Y" spends the night at his friends' houses and they spend the night at his house without a problem. (Arona Ragins, field notes, March 6, 1995)

Fatalistic attitudes receded when medicine provided concrete hope, assurances of its efficacy. Mr. T's fifteen-year old son thus became more optimistic about his future upon coming to the United States, where he saw for the first time that there were thalassemia patients older than thirty, leading him to believe he might conceivably grow up and even marry. Bone marrow transplants offered the greatest hope in that they represented permanent cure, obviating the need for transfusions. Mr. A was among such hopefuls:

> . . .my wife and I still hold the view that transfusion is not a long-term solution. Although it alleviates the symptoms of thalassemia, blood transfusion is not a solution, not a true solution. The basic disorder still persists. As she grows older,

> given the frequency of transfusion she will run into some point in time, some of the anti-genetic effect, having too many antigens in her body. That she will start rejecting transfusions. . .the bone marrow transplant. . .if it works, then she would haven't to do that again. . .
>
> (father, 43 years old, college-educated)

Comparing the cost of hospitalization for this procedure with the cost of regular transfusion and other related medicine, he estimated that with the bone marrow transplant, the family would "break even" in five years.

The expense and long wait for a bone marrow donor did not discourage poorer parents from this same hope since the ideal donor is a sibling. A 28-year-old mother, Mrs. Z indicated she had gambled on having a second child in the hopes of a match. She had a son, who turned out to be a carrier, not a match. However, her third child, a daughter, proved to be match.

A vigilant orientation also invited alternative perspectives, especially folk or popular healing approaches. These were not necessarily last-resort measures but parallel approaches—a finding consistent with other studies noting patients' co-use of both traditional medicine and conventional biomedicine (O'Connor 1995; Gesler 1991). A strong cultural tie to a distinct medical tradition may encourage this. Alternative approaches were often less expensive as well. Grandparents in Mrs. Z's household, for example, took their then two-month grandson to China to confirm the genetic diagnosis, consulting with a number of different doctors. Altogether, the child stayed in China for six months and was treated with a combination of acupuncture, more frequent transfusions, and iron pills. In this household of fourteen individuals, the total family income fell in the range of $16,000 to $29,999, yet the cost of treatment amounted to $50 in US dollars. Eventually, it was the low blood supply and the lack of individual medical attention (one nurse for every twenty children) that influenced the family's decision to bring the child back to the United States for treatment.

Some patients felt traditional foods or medicines helped their condition. In one Laotian family, these included "bitter foods," boiled tree roots. Others were more skeptical, such as Mr. A whose college background oriented him towards an extremely rational and intellectual approach that included reading a range of medical and technical articles, in Chinese-language sources as well as Eng-

lish. While traditional healing practices were investigated along with advances in Western medicine, the former were eventually terminated because progress could not be objectively measured. As he explained, the process was "to a large degree like rolling the dice, but you are in the blind. You don't know whether there's any progress made, or if there's any slip back being made, because there's absolutely no way of monitoring it."

Summary, Discussion, and Update

As recent immigrants, Asians have had a very different historical relationship with the medical establishment from whites or African Americans. It is basically their first encounter with the American health care system. Unlike families dealing with sickle cell or cystic fibrosis, who gave voice to the idea of adversity as "challenges" those affected by thalassemia held no such romantic notions. Many expressed the sentiment that had they *known* about the disease, they would have opted for abortion. Yet there were many social and cultural barriers to seeking knowledge, including fear of being rejected should they carry the genetic trait for thalassemia. Thus, even though prenatal diagnosis can theoretically prevent genetic diseases by giving couples the option of informed choice (e.g., selective rather than indiscriminate abortion) (Anionwu et al. 1988; Modell, 1990; Gamberini et al. 1991), procrastination and avoidance are common.

Cultural beliefs, in turn, encourage an attitude of acceptance, whatever one's fate. Toutu Vongphrachanh, a thalassemia outreach coordinator working mainly with Asian American communities, explained that Laotians view the birth of a child as a "blessing," a life to be nurtured and cared for. To abort invokes the fear of being "cursed" with bad luck (Vongphrachanh 2005) Thus, even if prenatal information had been available, abortion is not always a viable alternative. However, living with the disease is rarely *experienced* as a blessing. An eighteen-year-old Laotian patient, who is Buddhist, succinctly stated how she thought a child born with thalassemia would feel: "Bad luck. Painful. Wish they were never born." (Woo, field notes, September 21, 1993).

Despite the challenges of living with thalassemia, prevention is not without controversy. While public health screening is relatively uncontroversial for other diseases (e.g., syphilis, malaria, and tuberculosis), controversy has attended screening for genetic

diseases associated with specific ethnic and racial groups, given the political issue of who is screening whom, and for what purposes. Adult carrier screening determines whether a person has a certain genetic trait. However, Troy Duster, principal investigator of the gene screening project and a spokesperson on the moral-ethical issues of gene screening, observes that such knowledge has in the past led to stigmatization and discouraged patients from seeking counselling (Duster 2003). This is true even though carriers are generally asymptomatic and have no health risks. Negative public reaction, moreover, is more likely when a proposal for testing does not have the direct support of those communities or risk populations who will be affected. Indeed, any moves towards testing will likely be perceived as unwarranted efforts at "control" unless such programs are introduced as a result of the direct lobbying efforts of that specific interest group, as in the case of Jews at risk for Tay Sachs disease (Duster 2003). Secondary prevention through early detection of the disease in the newborn, however, is presently uncontroversial, as is tertiary prevention aimed at minimizing deterioration and complications among those already ill (e.g. through blood transfusions and Desferal treatment). Occurring as it does after the birth of the child, newborn screening does not involve highly charged decisions about whether to conceive or abort (Duster 2003). On the other hand, if screening is undertaken to *prevent* certain births, neutral attitudes are unlikely even though the test is a relatively inexpensive and simple procedure; the discovery of the trait, moreover, would also require a method of record-keeping and some kind of follow-up in terms of counseling or education (Duster 2005).

Since this study was completed, Dr. Vichinsky must be credited for his concerted efforts to deliver medical services in a more comprehensive way. Children's Hospital now provides an annual comprehensive checkup involving not only the hematologist but also many other specialists in the patient's care (e.g., endocrinologist, cardiologist, dietician, psychologist, social worker, etc.). The positive impact of this coordinating effort on a patient's life cannot be overstated: "All the people clear their calendar. . .so that everyone who is involved in that care is there. It's great because ideally it takes only two hours, whereas if I saw them one by one, I could be there from nine a.m. to two p.m." (Nguyen 2005). The pioneering nature of this radically different approach to thalassemia patient

care cannot be overemphasized, and is reflected in the collective efforts of those who worked to establish clinical guidelines for such comprehensive care in California (Quirolo et al. 2000). Another relatively new change implemented is the "family house," where patients can stay over. The hospital environment is a friendly and comfortable one where patients can "hang out and relax," participate in a patient community or support groups, rather than being simply a "filling station" for blood transfusions (Yamashita 2005).[3]

Problems of compliance persist. But the success of the program is evident in the fact that the patient population now includes a generation of young adult patients, an indication of the longer life expectancy that comes with better care. At the same time, this patient population includes many college kids who have adopted an avoidance strategy. "There are these smart kids, in college, working on a degree, whose health needs attention and who know this should be a priority and yet they're not doing it. . ..They know that Desferal is what keeps them alive and yet they don't do it" (Vongphrachanh 2005).

Recommendations

The patients who were pediatric patients ten years ago are now young adults—the first generation of Asian adult patients with thalassemia in the United States. Managing their illness in a pediatric facility is less than ideal, given certain social and economic issues specific to this age group. Some recommendations follow:

1. Learning to Tolerate and Work with Uncertainty

Although conventional Western medical practice is oriented primarily towards assessing a patient's objective condition, chronic conditions like thalassemia may have cultural or community connotations that burden the individual with a strong sense of self-blame. The language of "risky behaviors" can reinforce these feelings of guilt, and efficiency in the delivery of objective facts about a patient's condition can give rise to fears that undermine coping skills. Instead, communicating in ways that permit a certain amount of uncertainty may be preferable at times, if only because it permits the patient a certain sense of control, whether it is through an avoidance strategy that an individual adopts until he or she is emotionally ready to assimilate new information, or a vigilant one that leads the patient to adopt alternative approaches that

doctors might otherwise discourage. Medical staff who recognize this need will be able to work better with patients, and in the end will be more likely to establish the necessary rapport that enables the patient to become more involved in his or her own care.

2. Insurance Issues

The first major issue patients encounter when they approach adulthood is that they may no longer be covered by their parents' insurance. During the period of transition towards getting her own insurance coverage eight years ago, Laurice Levine paid $12,000 out of pocket due to a required eleven month wait period (Levine 2005). Financial considerations can therefore prevent patients from having access to the medications they need to be compliant.

Insurance issues also play a large role in whether patients have access to technology that facilitates compliance. Given general ignorance about thalassemia and the subtle stigma attached to being chronically ill, young adults do not want to appear different from their peers. Desferal treatment is generally bulky and conspicuous (e.g., beeping sound). A smaller, more easily hidden pump is generally not covered by insurance, unless it is private insurance. Yet such an alternative, Levine (2005) claims, "would promote compliance. . .They're great and most kids want to try this smaller pump."

3. Improving the Organization Of Care and Raising Professional Awareness

The single major issue that could improve the quality of patient life is the organization of services. There are still issues with patient wait-time that could be addressed. Wait-times were reported to be unnecessarily long—as much as half an hour even during periods when there are no other patients. Huythong Nguyen, a board member of the Thalassemia Action Group (TAG) and a patient since 1991, reported that admitting staff continue to either treat him as a "new patient" or assume he is a "parent," or else mistakenly try to redirect him to other hospitals serving adults. There is no institutional form of record-keeping that would eliminate these routine problems. The problems would no doubt be exacerbated for non-English speaking patients.

The problems are compounded when a patient with thalassemia seeks care elsewhere. Ironically, while the chronically ill are stereotyped as being helpless, the system of health care itself ren-

ders patients helpful. For one, the mere act of traveling or being mobile, which so many people take for granted, requires considerable forethought and planning.

> Your life revolves around *thal*. . .whether you're going on vacation, whether taking a quick weekend trip, going off to school. You gotta know when you're next transfusion appointment is. Where's a decent pharmacy where you can get your medicine? Where's a hospital that you can get to? . . . . (Nguyen 2005)

Second, many doctors themselves know little if anything about the disease. In the case of alpha thalassemia minor, for example, physicians have mistakenly prescribed iron supplements that have little effect on the anemia (Cooley's Anemia Foundation, 2001). This kind of ignorance can have critical consequences for adult patients shuttled from facility to facility because there is no resident expertise. Thalassemia is presently unknown to most medical doctors, and only to a select group of pediatric hematologists like Robert Yamashita, a researcher with the Cooley's Anemia Foundation and the Thalassemia Research Action Network. "Most hematologists do their primary work in oncology—why departments are "hem/onc"—working on such diseases as leukemia. So there are very few hematologists who know much about *thal*." (Yamashita 2005)

Laurice Levine and Huythong Nguyen both spoke poignantly of what it is like to seek health care under these conditions. "It's horrible. Nine out of 10 doctors—more than that—don't even know what thalassemia is. . .it's a nightmare trying to explain it to one doctor, then to another." Patients are thus hard pressed to find appropriate care at other facilities, including emergency rooms, where a patient might go for "respiratory distress, swelling, pain, or more commonly, a fever over 101" (Levine 2005). Nguyen (2005) adds, "We don't die from *thal* but from the complications."

4. Raising Public Awareness about the Disease

To enable individuals with thalassemia to reach a higher quality of life, outreach and education will need to occur on many levels, in many contexts. The social stigma associated with the disease is partly cultural (Choy et al. 2000), but it is also reflected in societal attitudes towards those who are different. Those attitudes, along with the institutional barriers to getting insurance, negotiating re-

lease time from school or work to obtain medical attention, can be harder to deal with than treatment itself.

> Social interventions would make a tremendous positive impact. The most challenging things for me as a patient (and I can speak for many) has been medical insurance, time off from school/job for blood transfusions, being served as an adult patient in a pediatric facility, and most of all societies views of chronic illness. These things are far more traumatic than getting blood transfusions every three weeks and poking myself with a needle nightly to do Desferal.
>
> You get reactions, like "I feel sorry for you. How can you do it? I could never do that." . . .There's a stigma, I'm sure. . .I couldn't even do PE (physical education). That's a stigma. Sometimes you look different. Your skin color might be different, or you have a large spleen. There are bone changes. . . . Or just having to miss school to get blood. . . There's falling behind in school. There are employment issues. For some cultures, marriagability is an issue. People often treat you like you are helpless when you are not. There's a stigma that if you have a chronic illness then you can't do things. (Levine 2005)

Outreach to the general public is therefore critical because of the possible impact on social attitudes within the community as well as on doctors and other health professionals who might encounter an individual with thalassemia. Health fairs held at college campuses would be a model venue. For one, a majority of this population is at an age when they are likely to be thinking about sex, reproduction, and marriage, even if some of these decisions may not occur until after they are settled in a career or longstanding relationship. I invited Vongphrachanh, Levine, and Nguyen to address forty-five students in my Asian American health class in the Fall of 2005. Students were not only forthcoming in their questions about the disease but expressed interest in getting genetically tested themselves and letting their friends and family know. Although the costs associated with genetic testing can themselves be prohibitive, campus testing can help save thousands of dollars in the long run. Counseling would also be more effective when integrated in an educational environment. Simply raising awareness about thalassemia could do much to dispel the ignorance. In the words of Levine (2005), "When people stop asking me what thalassemia is, then I'll know we're done with outreach."

References

Allen, S.J., A.O'Donnell, N.D.E. Alexander, M.P. Alpers, T.E.A. Peto, J. B. Clegg, and D.J. Weatherall. 1997. "*a*+-Thallassemia Protects Children against Disease Caused by Other Infections as well as Malaria." *Proceedings of the National Academy of Science U.S.A*. 94(26): 14736-14471.

Anderson, Helen M. and Helen M. Ranney. 1990. "Southeast Asian Immigrants: The New Thalassemias in Americans." *Seminars in Hematology* 27(3): 239-246.

Anionwu EN, N Patel, G. Kanji, H Renges, M. Brozovic. 1988. "Counselling for Prenatal Diagnosis of Sickle Cell Disease and Beta Thalassaemia Major: A Four Year Experience." *Journal of Medical Genetics* 25(11): 769-772

Association of Asian Pacific Community Health Organizations (AAPCHO) 1999 . *Genetics and the Primary Care Provider—with a Special Focus on Asian Americans and Pacific Islanders.* March.

Choy, Joanna, Dru Foote, Jennifer Bojanowski, Robert Yamashita, and Elliot Vichinsky. 2000. "Outreach Strategies for Southeast Asian communities: Experience, Practice, and Suggestions for Approaching Southeast Asian Immigrant and Refugee Communities to Provide Thalassemia Education and Trait Testing." *Journal of Pediatric Hematology Oncology*. 22(6): 588-592.

Conrad, Peter. 1994. "The Meaning of Medications: Another Look at Compliance," Pp. 149-161 in *The Sociology of Health and Illness: Critical Perspectives*, eds. Peter Conrad and Rochelle Kern. New York: St. Martin's Press

Cooley's Anemia Foundation. 2001. "What is Thalassemia Trait?" Flushing: Cooley's Anemia Foundation.

Duster, Troy. 2005. Interview by Deborah Woo. Berkeley, California, 27 August

Duster, Troy. 2003. *Backdoor to Eugenics*. New York/London: Routledge.

Duster, Troy and Diane Beeson. 1997. *Pathways and Barriers to Genetic Testing and Screening: Molecular Genetics Meets the "High-Risk Family." Final Report,* October.

Eleftheriou, Androulla. 2003. *About Thalassemia*. Nicosia, Cyprus: Thalassemia International Federation (TIF).

Gamberini MR, M. Lucci, C. Vullo, B. Anderson, R. Canella, I. Barrai. 1991. "Reproductive Behaviour of Families Segregating for Cooley's Anaemia Before and After the Availability of Prenatal Diagnosis." *Journal of Medical Genetics* 28(8): 523-539

Gesler, Wilbert M. 1991. *The Cultural Geography of Health Care*. Pittsburgh: University of Pittsburgh Press.

Heer, Nicole, Joanna Choy, and Elliott P. Vichinsky. 1998. "The Social

Impact of Migration on Disease: Cooley's Anemia, Thalassemia, and New Asian Immigrants." *Annals of the New York Academy of Sciences* 850: 509-511.

Lerner, Barron H. 2006. "Saying No is a Patient's Choice, However Risky." *The New York Times*, 11 April.

Levine, Laurice. 2005. Interview by Deborah Woo. Oakland, California, 22 August.

Lorey, Fred. 2000. "Asian Immigration and Public Health in California: Thalassemia in Newborns in California." *Pediatric Hematology Oncology* 22(6): 564-566

Modell Bernadette. 1990. "The Ethics Of Prenatal Diagnosis And Genetic Counseling." *World Health Forum* 11(2): 179-186.

Modell, Bernadette and Anver M. Kuliev. 1993. "A Scientific Basis for Cost-Benefit Analysis of Genetics Services." *Trends in Genetics* 9(2):46-52.

Nidorf, Jeanne F. and Kim-Yen Ngo. 1993. "Cultural and Psychosocial Considerations In Screening For Thalassemia in The Southeast-Asian Refugee Population." *American Journal Of Medical Genetics*, 46(4): 398-402.

Nguyen, Huythong. 2005. Interview by Deborah Woo. Berkeley, California. 2 September.

Northern California Comprehensive Thalassemia Center. 2003. "Thalassemia Trait." <http://www.thalassemia.com/thal_trait.html> as of August 25, 2006.

O'Connor, Bonnie Blair. 1995. *Healing Traditions: Alternative Medicine and the Health Professions*. Philadelphia: University of Pennsylvania Press.

Ostrowsky JT, A. Lippman, and C.R. Scriver. 1985. "Cost-benefit Analysis of a Thalassemia Disease Prevention Program." *American Journal of Public Health* 75(7): 732-736.

Parsons, Talcott. 1951. *The Social System*. New York: Free Press.

Quirolo, Keith, MD, Tom Coates, MD, Titi Singer, MD, Paul Harmzts, MD, Jorge Daaboul, MD, Vicent Gilsanz, MD, John Wood, MD, Mark Walters, MD, Neena Kapour, ME, Laurice Compagno, MA, CCLS, Rachelle Ang, MSW, Dru Foote, PNP, Deborah Harris, RN, MN, Susan Carson, RN, BSN, Elliott Vichinsky, MD. 2000. "Clinical Practice Guidelines For the Management of Thalassemia Patients: California Consensus." Proposal presented at Thalassemia Providers Conference. Pasadena, CA. June

Rund, Deborah, M.D. and Eliezer Rachmilewitz, M.D. 2005. "B-Thalassemia: Medical Progress." *New England Journal of Medicine* 353(11): 1134-1146.

Thalassemia Action Group. 2003. "Thalassemia Clinical Research Network." *Newsletter*.

Vongphrachanh, Toutu. 2005. Interview by Deborah Woo. Oakland, California, 22 August.

Vullo, Rino and Bernadette Modell. 1990. "What is Thalassemia?" New York: Cooley's Anemia Foundation, in conjunction with Thalassemia International Federation, with the cooperation of the World Health Organization.

Weissman, Joel S., PhD, Joseph Betancourt, MD, MPH, Eric G. Campbell, PhD, Elyse R. Park, PhD, Minah Kim, PhD, Brian Clarridge, PhD, David Blumenthal, MD, Karen C. Lee, MD, MPH, Angela W. Maina, BS. 2005. "Resident Physicians' Preparedness to Provide Cross-Cultural Care." *Journal of the American Medical Association* 294: 1058-1067.

Weitz, Rose. 1994. "Uncertainty and the Lives of Persons with AIDS." Pp. 138-149 in *The Sociology of Health and Illness: Critical Perspectives* eds. Peter Conrad and Rochelle Kern. New York: St. Martin's Press.

Yamashita, Bob. 2005. Interview by Deborah Woo. Berkeley/San Marcos, California. 19 August.

Notes

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1. This estimate includes physician and medication costs, with the possibility of exceeding this amount if there are medical complications (Toutu Vongphrachanh, Thalassemia Outreach Coordinator, August 11, 2005).
2. In one situation, where the father is an engineer, he is able to regularly take a vacation day once every five weeks to accompany his daughter for her transfusions. In turn, her absences from school are coordinated with the school calendar, and reciprocal arrangements are made with "study pals" to pick up homework. Both parent and child thereby minimize any disruption that would derive from missed work or missed school days. In another family situation, where the household includes an extended family of 14, there was a clear division of labor that developed around the care of a two-year-old with thalassemia. The twenty-eight year old mother had the main responsibility for bringing the child in for treatment, holding a daytime job as a bilingual telephone receptionist and a night-time job as a waitress. When she is at work, her father takes care of her two

boys, as well as cooks and cleans. Her mother, sisters, and nieces, also work as well as help with the child-rearing in various ways. In turn, the mother reciprocates by depositing paychecks, dividing the family income, and otherwise serving as the English-speaking go-between.

3. Some ambivalence attends the fact that some patients perceive the hospital as their "home," for it is indicative of the fact that their home lives are so wanting. "There's a kid who's twenty who has spent so much time here. He slipped and said, "I'm going home at 2:00, and he meant the hospital. He has no real home life or parents to guide him. Can't really even make a decision on his own" (Levine 2005).

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