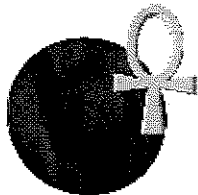


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## **Transition of Patients with Sickle Cell Disease from Pediatric to Adult Care**

Transition is a constant element of life. The process begins at birth and ends at death. Some transitions (for instance, becoming a parent) are more difficult than others (e.g., learning to drive). Some transitions, such as school graduations, are abrupt and marked by ceremony. Other transitions are gradual with less well-defined boundaries (becoming middle-aged, for instance).

People with chronic illness face the same transitions. Additional issues arise that most healthy people never encounter, however. Children with chronic illness that stretches into adulthood face the same challenges of adolescence as their healthy counterparts. The challenges are more complex and intertwined with other formidable tasks, such as finding new care providers and institutions. People with sickle cell disease often need help and guidance as they cross life's stream that separates childhood and adulthood.

### **The Issues**

#### **The Patients**

Success in the struggle to prevent childhood mortality in people with sickle cell disease raises new challenges for both health care providers and patients. Women need an array of services, including PAPP smears and mammograms. Both men and women face issues ranging from contraception and sexually transmitted disease to family planning. Young people with sickle cell disease must navigate through an increasingly complex medical care delivery system without guidance from parents or other adults. The transition from a pediatric to an adult care setting must take into account all these concerns and more.

Patients with sickle cell disease require coordinated care from subspecialists such as cardiologists, endocrinologists, ophthalmologists, gastroenterologists, surgeons, and pulmonologists. Transfer of information is a major problem for anyone with a chronic disease requiring care from multiple providers. This problem is compounded when care is transferred to a different setting as occurs in the shift from pediatric to adult clinics. Some of the subspecialty relationships play out over prolonged periods of time. For instance, endocrinologists treating delayed pubescence must coordinate activities closely with the hematology staff. Problems can develop if a patient's hematology care moves to a different setting while treatment is ongoing with key subspecialty providers.

The consequences of sickle cell disease vary tremendously between patients. Some 18 year-olds lead relatively normal lives, while others are virtually incapacitated by the disease. Complicated patients are most in need of smooth transition. Too often, however, precisely these patients face the greatest challenges with respect to transition. Platt et. al. (Platt, Thorington et al. 1991) showed in a prospective study of over 3,500 patients with sickle cell disease that frequency of pain episodes is a good clinical indicator of disease severity. For patients over age 20, higher frequency of pain episodes tended also to correlate with earlier mortality. Efficient transfer of these patients' care is critical to prevent undue morbidity and mortality.

An extensive study from the Duke Comprehensive Sickle Cell Center showed that after controlling for age and number of painful episodes, the patient's coping style significantly predicts health care contacts, activity, and psychological distress (Gil, Williams et al. 1991). The number of painful episodes correlated positively with age as well as with emergency room and physician office visits. Patients with Active Coping styles (use of multiple cognitive and behavioral strategies) had fewer emergency room visits. Those using Passive Adherence coping styles (reliance on concrete, passive approaches to pain, such as resting, with little flexibility when initial efforts fail to diminish pain) had more emergency room visits, and participated in fewer activities at home and in school.

Negative Thinking (self-statements of fear and anger) was positively correlated with psychological distress (Gil, Thompson, Keith, et. al., 1993). Parents' coping styles also correlate with children's activity level, distress and coping strategies. By following up on subjects 9 months after the initial assessment, these studies showed that while the coping strategies of younger children and adults are relatively stable, those of adolescents are in flux. For individuals who rely on less adaptive strategies (Passive Adherence or Negative Thinking), adolescence may be a period when these maladaptive styles become entrenched. The authors suggest (Gil, Thompson, Keith, et. al., 1993) "longer-term follow-up studies to trace the changes in coping over time and through transition periods from childhood to adolescence and from adolescence to adulthood". These stresses and changes complicate transition.

The unpredictability and stress of sickle cell crises affects all members of the family. The extent of the psychological impact reported varies between studies. Some show little psychological variation between adjustment in siblings, patients, and little difference from population norms (Gold, Treadwell et al. 1995) while others show significant stress on siblings (Treiber, Mabe et al. 1987). Kramer (Kramer and Nash 1995) found 49% of the parents of sickle cell patients in their sample to be clinically depressed according to the CES-D. Other programs have aimed at improving the education and coping of family members, reducing daily strain, and teaching stress management techniques to parents (Kaslow, Rowland et al. 1995), (Shearer, 1995). Maternal coping and family functioning correlate with patient adjustment and health care utilization (Shearer, 1995). Patients' self-esteem correlates with parental knowledge of sickle cell disease, more available social support for parents, and more adaptive parental coping strategies.

## **The Programs**

Social service providers facilitate patient/family adaptation to chronic illness. They are critically important when psychosocial issues complicate and threaten to impair medical management. Social service providers consequently may be the most important members of the transition team. They can help pave the way over the inevitable bumps involved in transition. Social service providers lay the groundwork for transition, and can intervene in problems before they escalate to crisis proportion.

The simmering revolution in health care and health coverage has largely overlooked the needs of patients with chronic illnesses. Few HMOs voluntarily roll patients with sickle cell disease beneath their umbrella. The challenges are even greater with for-profit enterprises. These organizations seek a patient portfolio weighted toward the young and healthy. Patients with chronic illnesses such as sickle cell disease frequently lose medical coverage when they become (legally) independent of their parents.

Integrated transition programs are critically needed to provide age-appropriate treatment plans, so that 30 year-old patients are not treated in pediatric clinics along side six-year olds. The psychosocial challenges of living with a chronic illness must be addressed if each person is to achieve his or her maximum potential irrespective of yardstick. Fear, depression and hopelessness can lead young people to maladaptive options. Equally importantly, society loses the latent talent in these young adults, which

is not fully tapped.

Parallel but unconnected health care systems for children and adults with sickle cell disease can lose patients during the transfer process. A number of factors conspire to this end. Perhaps most important is the attachment to a familiar institution and providers. Young people have eidetic memories of pediatric providers to whom they may literally owe their lives. They do not want to go to an adult care facility that they perceive as being cold, uncaring and unfamiliar.

On the adult side of the divide, patients face for the first time a plethora of new responsibilities, including keeping track of appointments and medications. Most young adults need keep track of nothing more serious than appointments for dental hygiene. Compliance failure can lead to caries. Young people with sickle cell disease must remember ophthalmology exams, with the penalty for non-compliance being possible blindness. In the absence of a transition bridge, young people with sickle cell disease often have difficulty crossing the divide.

## The Steps

A transition program must be tailored to fit the environment and available resources. Careful crafting is essential to avoid elements of the program existing in isolation or failing to intermesh with key counterparts.

### Assessment

All the interested parties, including providers, patients and institutions must agree on the definition of transition before any attempt at implementation. Transition is the orderly movement of the patient's medical home from one institution and set of providers to another. This contrasts with transfer, which is an abrupt and often unanticipated shift in locale. New providers can miss issues critical to the patient's health when presented merely with a recent discharge summary and a list of medications. Likewise, patients and parents must be properly prepared for transition if the process is to proceed smoothly.

Patient age should not be an automatic trigger to transition. Some patients will be ready for transition from pediatric to adult care by 18 years of age, while others will not. A patient's developmental age is a more appropriate guide than chronological age. Delay in neurocognitive development due to cerebrovascular injury, for instance, can mean that a patient may not be up to the challenges of an adult care setting until age 20 or 21. Similarly, a patient who has recently recovered from a serious complication of sickle cell disease, such as acute chest syndrome, is unlikely to be psychologically prepared for transition to new providers. Individual assessment is paramount.

Parents invest heavily in the care of their children. Sickle cell disease multiplies that investment and builds a tremendously strong bond between parents and pediatric providers. For parents, transition to adult care entails loss both of responsibility for and control over the young person's medical care. They often have difficulty relinquishing the central role in their child's care. The needs and concerns of parents must be assessed and addressed. The pediatric providers with whom they have a close relationship are positioned to do this best.

Patients and parents should view transition as a positive milestone. Together, they have achieved a significant step and should be congratulated. The pediatric providers should provide coaching and support for as long as possible after transition. The adult providers then become positive additions to an already winning team.

## Education

Both patients and providers must be educated about transition. The providers must structure the process to fit patient needs and institutional resources. Just as no physician can adequately evaluate chest pain without training in reading an EKG, no provider can construct an adequate transition plan with training in its complexities. Physicians, nurses and social service providers must work in close coordination to attain a smooth result.

Adult and pediatric providers must meet each other. This simple but crucial step is often difficult with the rising demands of clinical practice. Without this first step, however, attempts at transition are doomed. Pediatric providers are reluctant to send their patients into an unknown environment. This is true even if the recipient institution and providers have excellent reputations. Providers who have worked hard to bring children into adolescence and young adulthood are understandably cautious about delivering them to an ill-defined entity.

Regular meetings between adult and pediatric providers allow the groups to discuss the issues unique to each patient considered for transition. Medical records often do not contain important elements of a patient's history. Verbal communication and "group" knowledge of the patient often contain information that cannot be easily, if at all, converted to written format. Meetings assure pediatric providers that patient needs will be adequately addressed. These meetings also give adult providers a chance to ask for information they feel is important.

Successful transitions cannot be rushed. Both pediatric and adult providers must agree on the steps in the process. Furthermore, patients must be alerted to the impending changes as early as possible. The seeds of transition ideally are planted one or more years before the event. The steps should be delineated clearly both for patients and providers. The providers should assess success at each point along the path. Patients should have opportunities to ask questions and voice concerns.

The latter is particularly important. Patients sometimes feel as if their pediatric providers are abandoning them. This can produce both hurt feelings and resistance to the whole transition effort. Reassurance that the pediatric and adult providers will remain in contact is important to alleviating this concern. Patients who have information on transition months or years beforehand are less likely to resent the process.

## Preparation

Members of the adult provider team initially should meet the patient in the pediatric setting if possible. This provides a familiar, friendly setting for what could be an anonymous bureaucratic process. Ideally, this first meeting should occur prior to the final pediatric visit. The patient and parents can then process the information, and later discuss their feeling openly with their pediatric providers. The pediatric and adult teams can respond to issues in a familiar environment. New adult providers approaching difficult patient concerns in a foreign setting is a formula for failure.

When possible, a member of the adult team should escort the patient and parents to the new facility to familiarize them with the environment and staff. Finding a clinic a few blocks or even a few corridors away from one's home ground can be challenging. The issue is magnified for people with sickle cell disease, since they must often navigate their way while experiencing the agony of a vaso-occlusive pain crisis. A pain crisis is not an optimal setting in which to meet new providers.

The first clinic appointment is a crucial test of transition. Providers should not simply give patients the phone number and instruct them to make the call. At the last pediatric visit, the pediatric providers

should schedule the next appointment, just as they normally do. The appointment simply will be with the adult providers. If the patient misses the first appointment, a member of the pediatric team should phone to assess the situation. Often, patients remain reluctant about transition. They do not attend the adult clinic that they perceive as threatening. They do not return to the pediatric clinic, which they see as having abandoned them. The result can be loss of all medical follow-up. Patients often return in the throes of a medical catastrophe. Early intervention can avert disaster.

## **The Cast**

### **Social Service Providers**

Social service workers provide the proverbial glue for transition efforts. Patients and parents frequently have more contact with social service providers than any other members of the pediatric team. Many of the important issues of transition revolve around fear of the unknown. The effectiveness of the entire transition effort often depends on the handling of such issues. Each family is unique in its experiences and strategies of coping with the child's illness. Social service providers often have the broadest view of these concerns.

Following the introduction of transition as a goal by the health providers, the social service members of the team should begin to lay the groundwork. This includes discussions initially with the parents, and later with the adolescent. Reading material is an excellent way to reinforce the concept and serves as reference and reminder between clinic visits.

As the time for transition approaches, social service providers play a larger role. They often are best positioned to facilitate the introduction of the new adult care team. Patients and parents will have questions and concerns that can then be discussed and resolved. One of the most important tasks for the social service providers is to insure that the patient makes the first visit to the adult care clinic. If the appointment is missed, the social service provider often is the most appropriate person to phone the family and determine the nature of the problem. A "no show" at the adult facility may be an indirect way of asking for more help with the transition process.

### **Health Service Providers**

Physicians and nurses are the primary health service providers for most people with sickle cell disease. The organization of the care groups can differ significantly between pediatric and adult institutions. These differences can produce problems with transition unless they are recognized and properly addressed.

Primary pediatricians often are the axis of care for children with sickle cell disease. Hematologists generally are consultants. This structure arises from the fact that all children need pediatricians. The healthiest child makes several obligatory visits to the pediatrician for routine immunizations. Most preschools and day-care facilities require certification that children have received basic vaccinations, such as those for mumps, measles and pertussis. When added to common childhood illnesses such as chicken pox and Fifth Disease, pediatric primary care is the natural result. Since many children with sickle cell disease have relatively few additional problems, general pediatricians remain their focal contacts. Pediatric hematologists provide advice about specific interventions such as pneumococcal vaccination and penicillin prophylaxis.

By contrast, adolescents and young adults generally comprise the healthiest segment of the population. With the exception of required physical examinations for college or the military, few people between the

ages of 18 and 30 years have a standing relationship with health care providers. Visits to the doctor trail off with age, until only people with chronic ailments remain.

The result is that young people see doctors for specific problems, rather than disease prevention or health maintenance. The asthma specialist is the only medical contact for most young people with significant bronchospastic disease. The gastroenterologist is the primary provider for people with Crohn's disease. The hematologist is the primary provider for young people with sickle cell disease.

Young adults with mild asthma tend not to follow-up with pulmonologists. Only occasionally does this produce a serious outcome. Young adults with mild sickle cell disease tend not to follow-up with hematologists. Disasters can occur due to otherwise preventable complications of sickle cell disease. Retinal hemorrhage can produce severe and sometimes complete visual loss. The proliferation of retinal microvessels that precedes the catastrophe produces no symptoms. An ophthalmologist can detect and eliminate these vessels by laser surgery before complications develop. Preventative care is a key component in the management of patients with sickle cell disease. Young people who slip between the cracks during transition from pediatric to adult care often suffer unnecessarily.

Young people with sickle cell disease who remain in contact with pediatric providers can be transitioned to adult care more easily. These are often people who have suffered the greatest complications of sickle cell disease and are medically most complex. Adult providers sometimes feel they are given only the problem patients, while the pediatricians keep those who are easiest to treat. An organized transition program where the adult and pediatric providers meet to discuss the patients can dissipate ill feelings before they reify.

### **Patient Support Groups**

One of the most effective ways to diffuse fears of transition is to arrange meetings with people who have gone through the process. Patient support groups can be particularly helpful. Older adolescents and young adults reinforce the fact that youngsters have a future. Many youngsters and parents have the mistaken notion that sickle cell disease means a short, unproductive life. Teen support groups help to dispel the myth.

Patient support groups often meet in community settings. Patients and parents sometimes find frank discussion easier in the absence of health providers. Gatherings arranged around specific events, such as picnics or holiday celebrations, often work better than meetings focused solely on sickle cell disease. Patients often grow tired of reminders of disability. At a picnic, however, patients can discuss their concerns about sickle cell disease and transition. On the other hand, they can simply enjoy themselves in the company of other young people. Events are supportive. Meetings can carry an unfortunate sense of coercion.

### **Administration**

The administrators of the institutions that care for pediatric and adult patients with sickle cell disease are important to any effort to form a transition program. Some pediatric facilities have an upper age limit on people admitted for inpatient care. The work of the transition team is severely undermined if a patient deemed to need continued care by pediatricians, perhaps because of developmental delay, is transferred to the adult facility solely on the basis of age. The administrative personnel of the institutions must acknowledge the value of the transition teams and work with them to smooth transition efforts.

Transition programs require personnel. Administrators usually determine the personnel mix of the

institution. A single adult provider working without nursing or social service support cannot engage in transition activities. Even if the pediatric providers do the preparation, the effort will fail since a single overworked physician cannot adequately care for new patients. Transition is a program that exists between institutions. The world's most devoted providers cannot transition patients without support from those institutions.

## **The Challenges**

Committed personnel are key to the transition effort. The infrastructure of the medical care system greatly influences the structure of the program and the obstacles that must be overcome.

### **Parallel and Equal Programs**

Coexisting, well-developed programs for children and adults with sickle cell disease provide the simplest setting in which to develop a transition program. Their existence does not guarantee effective transition, however. The elements previously reviewed must be worked through in the best of settings. The social service providers are the key link between the adult and pediatric programs. They require support from the medical staff and the institutional administrators to bring the effort to fruition. In this setting, patients with sickle cell disease can grow into adulthood without discontinuities in their care.

### **Parallel and Unequal Programs**

Pediatric and adult programs of unequal strength challenge the concept of transition. The most common scenario in the United States is a well-functioning pediatric program with an adult program that is nonexistent or rudimentary at best. A number of factors conspire to this end. Most children with sickle cell disease are offspring of heterozygous parents. The child is covered under the parent's health insurance, allowing adequate compensation to the pediatric providers and institutions. Furthermore, children with sickle cell disease often have fewer chronic complications than do adults. Acute pain crises are both the most common cause of hospitalization in children, and are self-limited.

In contrast, adults with sickle cell disease frequently have difficulty maintaining steady employment in the absence of flexible jobs that take their disability into account. The consequence is that a disproportionate number of adults rely on assistance from the government, including mandated health programs. In the United States, these programs often fall short of comprehensive coverage. Adults with sickle cell disease, like other people with chronic illness, lack the breadth of services that would support activities such as transition programs.

Chronic end-organ damage begins to manifest in adulthood. Problems such as osteonecrosis, renal dysfunction, and cardiac dysfunction are more frequent in adults than in children. The combination of chronic, intractable problems, such as osteonecrosis of the femur, and poor financial compensation deters some adult providers from the care of patients with sickle cell disease. The advent of managed health care has exacerbated the problem, since these patients are least attractive to people who focus on profit/loss margins.

The result is that many pediatric programs lack an adult counterpart to which their patients can be transitioned. Pediatricians heroically hold on as their patients age and sometimes develop problems outside their area of expertise. A pediatrician learning to use nitroglycerin for patients with ischemic heart disease is an unfortunate testimonial to the often-dire nature of the situation.

The institutional commitment, particularly on the adult side, comes to the fore in this circumstance. Current data indicate that a well-organized adult care program for patients with sickle cell disease will be a financial plus for the institution (Benjamin, et al., 2000). There must be a cadre of patient advocates, both professional and lay, who hammer this point home. Patients with sickle cell disease often slip between the cracks when large institutions balance the competing demands for care presented by its diverse patient population.

Without a structured adult care program, pediatric providers can only equip their patients to find their way through the maze on the other side of the divide. Patients should learn to keep copies of their basic records, such as discharge summaries. They should also keep a list of current medications, including the medications that work for them during an acute vaso-occlusive pain crisis. Patients should keep a diary of important events such as immunizations and complications of their sickle cell disease. Finally, the pediatric providers should dictate a brief overview of the patient's medical course during childhood. This means distilling the data in volumes of charts to a couple of pages. The familiarity that the pediatrician has with the patient over the years makes tractable an otherwise formidable task.

## The Last Words

Transition programs prepare an adolescent to assume responsibility for his or her health care. As such, the primary charge lies with the pediatricians and parents. These two groups have invested most heavily in the care of the child. They naturally desire that the adolescent enters young adulthood in the most positive fashion.

Transition programs do not evolve naturally. Someone perceives the need and acts to fill the void. Most often, the impetus comes from pediatricians and parents. Adult providers sometimes are reluctant to engage in a transition effort, for the reasons outlined earlier. The initial phase that follows recognition of need is crucial to the ultimate outcome. Misstep at the outset can result in the erection of an iron curtain, instead of a transition program.

The components needed to establish a transition program vary tremendously. Attempts to establish programs must be individualized and account for differences between institutions, providers and patients. An assessment of needs is a key first step. Before building a house, the planner assesses the needs of the people who will occupy the structure. She also surveys the ground and terrain to determine the type of structure the environment would best support. Only then does the planner move forward with design and construction. The same is true for transition programs. An ounce of preparation is worth more than a pound of repair.

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