

# Chapter 18: Psychosocial Issues

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## Introduction

Each family confronted with a diagnosis of Fanconi anemia (FA) faces challenges in responding to the illness, from coping with the emotional upheaval of the news to assuming the responsibility of organizing their child's care. A diagnosis of FA often changes the family system, requiring parents to make important healthcare decisions that involve a sophisticated understanding of a complex illness with many treatment options. While any serious illness in childhood can isolate a family from their community, isolation is more likely with a rare condition such as FA. The challenge is for family members to balance their emotions while orchestrating their child's medical care, surrounding themselves with a community of support, maintaining hope, and sustaining some semblance of a normal family life.

Fanconi anemia presents different issues for families depending on the developmental stage of the patient and the patient's individual illness progression. The number of children with FA, the number of unaffected siblings, and their ages will also affect the emotions and needs of each family. This chapter will describe the stages of families' journeys with Fanconi anemia, as well as the unique challenges faced by parents, siblings, partners, and children, adolescents, and adults with FA. This chapter will also discuss stage-related psychosocial issues surrounding hematopoietic stem cell transplantation (HSCT) and end-of-life.

## Stages of the Journey with FA

Living with uncertainty and preparing for a future filled with the medical complexities of FA, while helping children embrace life and establish dreams, career paths, future plans, and hope for longevity places affected families on a unique and challenging journey. This journey can be interwoven with grief, loss, and uncertainty, but it is important for families to note that the course of FA is constantly changing, and that there is ever-increasing room for optimism after the impact of the initial diagnosis fades.

Before FA has been confirmed, most families remain hopeful that this possible diagnosis will be proven wrong. The time of diagnosis itself is an emotional crisis, a period of transition. It takes time before parents can move from shock and disbelief to a more proactive mode of coping.

At the time of diagnosis or soon thereafter, parents of children with FA may begin to face difficult decisions regarding medications or other treatment options. Families need access to up-to-date, clearly presented information to help them navigate this complex illness and make decisions with which they will feel comfortable. They may also need help thinking through their choices and the implications of those choices. Parents, in particular, face a number of uncertainties at the time of diagnosis: Will their child eventually need a stem cell transplant, perhaps from a human leukocyte antigen (HLA)-matched sibling <sup>(1)</sup>? Will they arrange for prenatal diagnosis or HLA typing with subsequent pregnancies?

Once the diagnosis has been established, many families find that emotionally calmer times alternate with more complicated ones. Children with FA can be stable or asymptomatic for long periods of time. As initially described in *The Damocles Syndrome* <sup>(2)</sup>, many parents feel as though they are constantly waiting for the next crisis <sup>(3)</sup>. Families may need help cultivating the ability to live each day to the fullest. Honing this ability is essential, as learning to focus on activities apart from the illness is an effective day-to-day coping strategy. Moments not driven by medical crises are times for families to enjoy life, prepare for the future, and stay abreast of salient treatment options. The information amassed during calmer times will help families feel prepared should their child's condition deteriorate and additional treatment options need to be considered. Education, and a strong support network empower family members to move forward with necessary tasks during times of emotional crisis, when feelings of hopelessness and immobilization may prevail.

For many of the difficult medical choices that face parents and their children with FA, there is no turning back. Therefore, major decisions require that families and older patients know all they can prior to moving forward. Not only should families take time to learn about treatment options, they should also have ample opportunity to integrate the information, and reflect upon and accept the choices they have made. In certain cases, families must make decisions about experimental procedures and protocols. Families may experience vulnerability and anxiety when they know they are traveling on a road that few have traveled before.

## The Parent's Journey with FA

### Coping strategies

Within each family, parents may cope separately and very differently with the diagnosis and course of FA. One parent may prefer to learn as much as possible to create a strategic plan for the future, while the other parent may prefer to focus on each moment. One parent may need to talk and to cry; another parent may be uncomfortable with displays of emotion. Differences in coping styles should be recognized so that each parent can be supported for his or her strengths, insight, and abilities during the course of the illness. Coping differences can create or aggravate marital tension, particularly in relationships that were already stressed before the diagnosis of FA. Alternatively, some couples feel that the magnitude of the illness has helped them forge stronger relationships.

### What Parents Can Do

- Recognize that there is much more to your child—and your family—than FA. Try to enjoy “normal” family activities, encourage your child’s dreams for the future, honor other siblings’ emotional needs, and celebrate each triumph no matter how small.
- Join FARF's online support groups for FA.
- Attend events sponsored by FARF and other organizations.
- Become an expert about FA, its treatments, and options for the future, such as preimplantation genetic diagnosis (PGD).
- Familiarize yourself with techniques for parenting children with chronic illness.
- Forge strong, collaborative relationships with experienced physicians.
- Learn how to explain FA to inexperienced care providers, educators, and others while respecting your family’s need for privacy.
- Advocate for your child’s best interests in education, health care, and elsewhere.
- Recognize the signs of depression and anxiety, and seek out emotional support when you or your family members need it.

Many parents living with FA feel anxious or depressed from the onset, unsure of what to anticipate. The abilities to manage these emotions, make decisions, continue to function, and enjoy life may not be present initially, but are skills

to be mastered as time goes on. Psychosocial support services can greatly assist families who find it difficult to function in the face of their emotional responses; it is important to encourage parents to seek help when they recognize that they need it.

Staying abreast of the ever-growing body of knowledge about FA and its potential treatments can help parents feel calm, focused, and grounded. Talking to other parents, understanding the processes of decision making, and getting support can help parents maintain the emotional balance they need. Information and support are readily available from the Fanconi Anemia Research Fund (FARF) through its website, Facebook pages, e-group, family materials, and communication with FARF staff. These resources support the ongoing and changing needs of children, adolescents, young adults, and adults with FA, and their families. For several years, the FARF has held its annual Family Meeting at Camp Sunshine at Sebago Lake in Casco, Maine (<http://www.campsunshine.org>). The meeting blends educational sessions, presentations about current research, psychosocial support, and recreation.

Support groups offer parents the opportunity to be parents: to compare their child with other children, to seek companionship of other parents in similar situations, to share information, and to join the fight against FA and become empowered in the face of the illness. Facebook, CaringBridge sites, CarePages, and other social media create a global connection among families affected by FA. At times, individuals may feel overwhelmed by the emotional commitment to others and excessive amounts of time spent on the Internet; when this happens, it is important to set personal limits and take a break if needed.

Parents may be incorrectly perceived as aggressive when they advocate for the best interests of their children. There may be moments when families and individual physicians do not agree on treatment options or alternatives (e.g., hematopoietic stem cell transplantation, the use of androgens, or other therapies). The professionals involved in the patient's care must work to make the best decisions *with*, rather than *for*, families. This strategy will help reduce the possibility of future regrets for families and professional staff.

Relationships forged with physicians can be of tremendous value and significance to families affected by FA. The quality of these relationships often influences the family's entire experience of the disease. By helping families navigate the course of the illness and think through decisions, physicians can help those living with FA feel part of a larger system rather than feeling isolated and alone.

Parents, caregivers, and other family members truly become experts about FA. They must integrate tremendous amounts of information while attending to their child's medical needs and managing all other activities of the family. It is not surprising that when parents of children with FA are asked what they have learned about themselves or their children since the diagnosis, they overwhelmingly affirm that they have learned how strong and capable they and their children are <sup>(4)</sup>. Parents describe having a greater appreciation for the things that they do with their children, and they often describe a newfound ability to experience each day to its fullest.

### **Parenting in a family with FA**

When a child is diagnosed with a life-threatening condition, concerns about the child's future often change regular parenting habits and instincts. These changes can profoundly affect the parent-child relationship, and can go on indefinitely in the case of a long-term illness like FA. Parents may turn to physicians for support in returning to normal parenting patterns once the crisis of diagnosis has passed; physicians can also provide help when a child begins to act out and display symptoms of externalizing behaviors, such as tantrums or rebelliousness. Limit-setting and structure make children feel cared for, safe, and secure. Excessive permissiveness by parents who are fearful or sad—or disconnected and inconsistent parenting by those who suffer from depression—unwittingly communicates to children that their behavior doesn't matter.

### **Young Adults' Advice to Parents**

- Don't waste time worrying about what will happen in the future. If it's going to happen anyway, there's no sense in worrying about it right now.
- Don't feel guilty or responsible for the disease. You are not to blame!
- Don't be overprotective.
- Don't forget that my siblings need your attention and support, too.

Parenting siblings without FA—and deciding whether to conceive additional children as potential stem cell donors—can be emotionally challenging as well. Through a process known as preimplantation genetic diagnosis (PGD), parents can determine the genetic makeup of an embryo before it is implanted through in vitro fertilization. With the recent refinements in this process, many families will try to have a child free of FA whose HLA-matched stem cells can be used in a transplant for their child with FA. This process can be financially,

emotionally, and physically draining and, in some cases, all-consuming. Unsuccessful attempts at PGD are disappointing and can create other conflicts for the family, as treatment options as well as additional children stand in the balance. Successful PGD attempts, joyous in nature, create an unusual dichotomy in which the family anticipates the transplant and the birth of a child simultaneously. Discussing concerns with others who have attempted PGD can help families mitigate the intense emotions that can occur during this time. Parents who are beyond childbearing age and unable to benefit from PGD may experience remorse that this technology was not perfected earlier in the course of their child's illness.

## **Siblings in a family with FA**

### **Unaffected siblings of a child with FA**

Siblings care and worry about each other a great deal and, for many children, the universe is defined by their role as either an older or younger brother or sister. The siblings of a child with FA experience their own unique concerns, some visible to other people and some invisible. They may feel guilty that the disorder happened to their sibling and not to them, or they may feel that they are less important, because they are not getting as much attention. Siblings of children with life-threatening illnesses often have as much of an emotional response to the illness as the affected sibling.

### **What Siblings Can Do**

- Ask questions when you don't understand.
- Set aside one-on-one time with your parents.
- Recognize that you are unique, important, and treasured in your parents' eyes—even if they seem distracted at times.
- If you so desire, ask how you can help and become more involved in your sibling's care.
- Learn how to explain FA to curious peers while respecting your need for privacy.
- Recognize when you or others feel sad, frightened, or confused. Tell someone how you feel!

Sadness, anxiety, jealousy, and guilt are common emotions experienced by siblings <sup>(5)</sup>. Providing opportunities to express these emotions, keeping the

lines of communication open, and learning how to process the experience can help siblings work through their emotional responses and find their place in the family system. It is important for families to address their unaffected children's feelings and questions. Siblings are best able to thrive when they can spend quality time alone with their parents, when they are provided with developmentally appropriate medical knowledge, and when they truly feel that they are an integral part of the family <sup>(6)</sup>.

Siblings benefit from having a consistent, designated caregiver in their lives, particularly during times when their sibling with FA is hospitalized. Perceptions of the medical care required by their siblings may be more frightening or more idealized than reality. Inviting the sibling to be a part of the hospital routine can be helpful, and can assist the sibling's ability to cope with the situation.

Unaffected siblings of children with FA will usually be tested for their suitability as donors should transplant become a possibility. Families must make every attempt to appreciate the emotional journey of the sibling donor, a journey that can be markedly different if the sibling is a match or not, or if the transplant is successful or not. Age-appropriate information and emotional support are essential throughout the process. Stem cell donors have their own experiences, which need to be heard and acknowledged.

### **Multiple children with FA**

The already complex relationships of siblings are further complicated when more than one child in the family has been diagnosed with FA. The experience of each affected child will have its own impact on the other affected children. Non-affected siblings may be carriers of FA which creates yet another, often unspoken, dimension in family relationships, especially when they reach child-bearing age. It is important that affected and non-affected siblings have opportunities to talk with each other and with their parents. Sibling relationships can be among the strongest in life and need to be cultivated and nurtured.

## The Child's Journey with FA

### Explaining FA to a child

#### What a Child with FA Can Do

- Ask questions when you don't understand.
- Get involved in activities that you enjoy.
- Recognize that you are unique, important, and treasured—and remember that there's much more to you than FA.
- Learn how to explain FA to curious peers while respecting your need for privacy.
- If you feel ready, ask how you can become more independent and involved in your own health care.
- Recognize when you or others feel sad, frightened, or confused. Tell someone how you feel!

How parents accept and face the illness will influence how children with FA grow and adapt. If parents create an environment that allows for questions, discussions, and an expression of feelings, children will feel free to ask them for information about their illness and treatment options, and become active participants in their own disease management <sup>(7)</sup>.

Children often know much more about what is happening than adults might believe. In addition to what they have been told, children pick up information from ambient conversations, have independent interactions with professionals, and surmise things from the emotional climate around them. Children will ask questions when they want to know about a particular issue, but will often shy away from questions to which they do not want the answers or to which they have not gotten responses in the past. Children are good regulators of their own knowledge base, providing cues to the adults around them at all junctures. Once children are able to read and have access to the Internet, they often perform online searches about their illness.

A major concern of parents is how and when to tell children about FA. At each stage of development, children need age-appropriate explanations of their diagnosis and treatment. These explanations should grow in sophistication as the child grows. Information offered regularly to children will enhance their ability to understand their disease and establish trusting relationships. As they



get older and medical problems emerge, groundwork set in earlier years will encourage affected individuals to rely on health care providers for answers and advice. Providing children with practical knowledge about FA can help them understand what is going on in their bodies, and why a certain treatment or medical test is needed. This information builds trust and engages children as active participants in their own care.

### **School-related concerns**

School is a powerful normalizing environment for children. Learning has been called the “work” of childhood, and brings structure and meaning to children’s lives. Supportive educational environments can make a major difference in a child’s quality of life by building confidence and hope as skills are improved in cognitive, social, and emotional domains. When school attendance is interrupted, or a child’s performance at school is impaired, it can be very disorienting and disruptive. Prioritizing school for children with FA, when medically safe, helps parents and children maintain structure and hope.

Children with FA may face unique challenges in school. Some may have cognitive impairments that require special attention. Others may have no known problems, but may need extra assistance because of illness-related absences. Others may have physical limitations and may need extra support. School is often where children with FA may begin to feel as though they are different from others, whether their differences stem from frequent absences, inability to participate in activities, or other perceived differences. Children may need help learning how to adapt, respond, and connect with their peers.

Visible characteristics of FA, such as short stature or missing thumbs, serve as constant reminders to the outside world that a child with FA is different. At all ages, physical and other differences may set children with FA apart from their peers and cause them to feel anxious, isolated, or depressed. These emotions can affect their self-esteem and their ability to focus on age-appropriate achievements. Children need to be able to confide in their parents and others when they feel physically or socially limited by FA; counseling may be of great benefit during these times.

Fanconi anemia is commonly associated with a range of neurological and developmental issues characterized by mild to significant impairment, such as attention deficit hyperactivity disorder (ADHD), learning disabilities, and developmental delay <sup>(8)</sup>. In addition, treatments for FA, such as anabolic steroids and HSCT, may affect cognition, mood, and behavior. Any child

with FA who is having learning or behavioral issues at school should be formally assessed with an individualized educational plan (IEP) through the public educational system. Treatment centers often provide comprehensive assessments and specific academic recommendations that teachers and administrators at school can follow to personalize the child's educational plan. Children typically struggle in school after returning from HSCT, and transplant teams should help patients and families navigate the special education system and any other resources needed to optimize the child's adjustment and success. Social workers, case managers, child psychiatrists, psychologists, and neuropsychologists can help families advocate for their children.

### **Growing up with FA**

School-age children develop increasingly strong relationships with their peers as they begin to differentiate themselves from their families. Physical limitations that require children to remain dependent on their parents may influence the extent of their social activities. Each child and family must learn to strike a balance in social and family relationships that allows for a blend of independence and dependence, nurturing and differentiation.

Children with FA are invariably exposed to difficult experiences due to the nature of the illness. They face multiple hospitalizations and medical treatments, and may be exposed to the deaths of siblings or other children with FA. These children may therefore come to understand and deal with issues of mortality with which adults may not feel entirely comfortable. Although parents may work hard to "normalize" their children's lives, patients with FA have unique experiences and are confronted with the concept of death at earlier ages than other children. Thus, children with FA often seem more mature than expected for age, and often have more sophisticated attitudes than their peers regarding matters of illness and death. These children may also appreciate life more than others they encounter. However, some children experience a disconnection between what they understand and how to cope with what they experience. An environment of active support and open discussion is helpful for children, but can become complicated if adults do not recognize the need for such discussions.

During adolescence, challenging the rules is age-appropriate and, at times, promotes emotional growth. It allows teens to assert themselves as individuals and begin learning how to take responsibility for their actions. For adolescents with FA, however, this can be a time of rebelling against the "rules" of the disease. Young adults sometimes stop taking their medications and migrate to

activities that have been discouraged, such as sun bathing, drinking alcohol, and smoking. Adherence to medication regimens is a serious concern and should be given particular attention at this developmental phase, as should behaviors that increase the risks of cancer. For adolescents who may already feel socially isolated, foregoing of age-appropriate, yet maladaptive behaviors may pose additional psychosocial issues. During the adolescent years, association with peer groups of others with FA can combat countercultural behaviors, as can the wisdom of adults with FA—particularly if they can recall and share their own adolescent experiences.

As children with FA get older, they should become actively involved in assenting, consenting, and participating in decisions about their medical care. During this transition, parents may feel some relief that they are now making decisions *with*, rather than *for*, their children. Nevertheless, many parents have expressed anxiety about their children's abilities to make complicated and important decisions for themselves as they enter young adulthood. For some young adults, medical decisions will continue to be made in collaboration with their parents; in some cases, cognitive factors may limit the young adult's ability to make decisions. Other young adults will desire full responsibility. When this happens, parents must begin to trust the choices of their grown children. This period of growth for the person with FA also becomes a time of growth for parents, and occasionally creates dissonance between parents and their children.

Living with FA can be a long and arduous journey for many children. Room for continued growth, regardless of medical issues, is a vital part of childhood and prepares children to be successful and motivated in life. Empowering the child with FA inherently helps all family members acknowledge and delight in the child's gains, rather than focusing exclusively on FA. Celebrating achievements—great and small—cultivates growth and satisfaction for both children and their parents, and reminds families that FA is a component of their children's lives, but not what defines them.

## Young Adults and Adults with FA

There is a large and ever-growing population of young adults and adults with FA. Adult FA patients serve as an inspiration to all, yet should be recognized for their own needs, aspirations, and struggles. The medical course of FA is evolving, allowing for a better understanding of emotional and physical

challenges. Emotional connections for this group can also be found in young adults and adults with other rare illnesses who have survived to adulthood.

The transition to young adulthood engenders a more comprehensive understanding of the illness, sometimes producing a new emotional response to FA. During this time, adolescents begin to address salient issues that may have lain dormant during earlier developmental stages. Young adults with FA begin to find their own voices, assume responsibility for managing their own illness, become the primary decision-makers in their care, view their parents as partners or consultants, and truly become independent—all very appropriate and significant steps. It is important to help young adults gain their independence while also letting them know that they can continue to rely on their families for support and assistance. Those who face more severe manifestations of the illness may, of necessity, remain more physically and emotionally dependent on family members. Many young FA adults find that their family connections are stronger than those of their healthy peers. Dependence on parental care and transition to independent ownership of health care can be a major source of empowerment and anxiety. It is best effected over time and with the guidance of adult caregivers.

People with FA approach adulthood from a rather unique perspective, having grown up with uncertainty about the future. These adults face the “normal” challenges of establishing and mastering life goals and forging lifelong commitments, but must also find ways to address the impact of FA on issues surrounding partnership, sexuality, marriage, children, ongoing cancer risks, financial issues, and concerns over medical insurance.

### **Staying on track**

Fanconi anemia affects the whole family—current and future generations—and not just when a child is first diagnosed, but throughout the course of the illness. Ideally, a strong, collaborative relationship between parents and children should be well established long before the transition to young adulthood. Family members should start building this relationship as early as possible by working together to adopt the best decision-making practices for their particular situation.

Ambivalence and anxiety can plague young adults with FA, who need to adhere to unique challenges of living with the illness while struggling to be like everyone else. Normal developmental challenges do not evade young adults with FA, yet age-appropriate experiences may have greater intensity and significance. Relationships, peer pressure, experimentation with drugs

and alcohol, and sexual relationships all pose unique emotional and physical challenges for young adults with FA. Given the inherent increased risks of cancer from a number of these behaviors, young adults with FA are torn between the desire to take care of themselves and the desire to enjoy typical age-appropriate experiences with peers.

Omnipresent are feelings of isolation and distance from adults who are unaffected by FA; thus, relationships among adults with FA often engender an unequalled sense of personal connection. It is easy to see how the multitude of illness-related factors might affect the day-to-day emotional well being of young adults with FA. Beyond the personal aspects of dealing with the disease, these patients may feel accountable for their behavior in the eyes of their peers with FA, parents, physicians, and other professionals with whom they have developed connections. This sense of accountability may encourage young adults to do the “right thing.” Although such connections may increase compliant behavior, online networks can also begin to resemble more typical peer forums that increase the desire to have more “routine,” but risky, social experiences, which ultimately challenge adherence to the patient’s healthcare regimen. Special care should be taken by members of the young adult’s support group to approach these issues with a caring, nonjudgmental stance, understanding that while these behaviors may increase the likelihood of cancer significantly, they are also complex and may have roots in a variety of areas related to family dynamics, desire to have a “normal” life, desire to escape thoughts of FA, or simply lack of understanding about the impact of these behaviors on personal health.

### What Romantic Partners Can Do

- Learn about FA: the causes, treatments, implications for the future, and preventative health strategies.
- Consider joining online support groups.
- Recognize and remember that there is much more to your partner than FA. Focus on activities that you both enjoy!
- Ask your partner if, when, and how you might become involved in his or her care.
- Learn how to explain FA to others while respecting your partner’s need for privacy.
- Recognize the signs of depression and anxiety, and seek out emotional support when you or your family members need it.

### **Dating and relationships**

Deciding when to tell potential romantic partners about FA is an integral part of the dating process for any adult with FA. The issues of *whom* to tell, *when* to tell, and *what* to tell are inextricably tied with concerns about whom to trust and how a relationship might be affected. These issues can silently frame the early stages of relationships with roommates and romantic partners. An open and honest parent-child relationship provides the patient with a model of communication that can help young adults feel comfortable informing others about FA without shame or fear.

As relationships flourish, there is a natural inclination to think about the future. This reflective process evolves as young adults ponder their future goals in the context of what they know about their medical prognosis. All of this may influence their choices in friends, relationships, careers, marriage, and parenthood.

Partners of young adults with FA often need help understanding the disease and its implications for their relationships, as well as the roles of other family members. Partners also need an outlet for information, expression, and support when their partner is not doing well or has to make major life decisions. Many partners understand the disease intellectually, but aren't able to articulate their own concerns until medical concerns arise. Negotiating their roles as partners, particularly when a patient's parents may have nurtured the patient for decades, can be quite challenging. Information, support, and counseling are important tools to help partners navigate this complex journey.

## **Coping with Stem Cell Transplantation**

Stem cell transplantation can be a turning point in the lives of individuals with FA and their families. This procedure may cure the hematological problems of FA, but it also carries risk of illness or death. The perception of HSCT has changed significantly as the outcomes of transplant have improved in recent years. Many families talk about HSCT as less a question of "if," but more often a question of "when." Waiting with uncertainty for long periods of time can be stressful.

### **Preparing for transplant**

Families enter into transplant from a variety of different perspectives. Some patients require HSCT soon after a diagnosis of FA in the context of acute hematologic complications, while others are able to plan in advance, and still

others face the prospect of a transplant in the undefined future. Psychosocial needs differ depending on the patient's perspective and factors related to their family: Are the parents single or partnered? Do other siblings have FA? Are alternate caregivers available? Do the parents have flexible work schedules? How far must they travel to the transplant center? Is the family financially secure?

Many parents experience anxiety, depression, and psychological trauma during the time of transplant. It is critical that they have ongoing access to practical and psychological support from social workers, psychologists, and psychiatrists during and after HSCT. Transplant teams typically include professionals skilled at helping families navigate various challenges, including medical leave from work, childcare for siblings, absence from school, and the practical medical care of their child.

Children of all ages should be prepared for HSCT by their parents, who can work with experienced pediatric professionals to provide explanations appropriate for each child's developmental stage and medical situation. Child life specialists are professionals trained in using play, education, and art to support children in the healthcare setting. Parents often struggle with knowing how much information to share with their children out of a wish to protect them from worry or fear. Children who can communicate openly with their parents about their illness feel safer and experience less anxiety and depression, no matter what their medical prognosis is. Children who have been appropriately prepared tend to cope better with treatment demands and symptoms, and most importantly continue to trust their parents as sources of information and support through hard times <sup>(9)</sup>.

### **Course of transplant**

The days and weeks prior to transplant and through the process of induction chemotherapy are usually the most anxiety-provoking for patients and families. Anxiety tends to peak at this time and decline significantly after the actual transplant infusion, even though patients may remain in isolation awaiting engraftment for several more weeks. The prolonged hospitalization can be marked by significant discomfort and symptoms of nausea, pain, and fatigue, which should be treated aggressively with medication. Patients often find relief in other modalities as well, including hypnosis, behavioral therapy, and relaxation techniques. For many patients, physical symptoms steadily improve over time but may persist past discharge into the outpatient setting, which can be dismaying to patients who have not been adequately prepared. Boredom

commonly causes distress during this time and should be staved off by crafting daily routines with the help of calendars, including structured sessions with physical and occupational therapists, school teachers (as appropriate for the patient's age and time of year), and other supportive members of the multidisciplinary team as well as visits from family and friends. This is the “work” of getting better, and patients need encouragement to stay active in their own recovery. During this period, caregivers need significant support coping with, and keeping track of, extremely complex medical regimens—in some cases, patients take as many as 20-30 medications daily.

Once the patient has been cleared to return to school, children should be involved in discussions and decisions about the re-entry process. School staff can help students by role-playing examples of conversations that might occur on the student's first days back to school. This exercise can help the student find comfortable explanations that balance his or her need for privacy with answers that will satisfy the curiosity of peers. Adolescents and young adults may struggle with adherence to the recovery plan in the post-transplant phase due to a variety of psychosocial factors, including denial, anxiety, developmentally typical struggles with dependency and vulnerability, or post-traumatic stress disorder. Patients should be assessed for these and other issues, and to encourage positive personal care behaviors.

## **Issues Surrounding Death**

When nearing death, the patient and the family need emotional support, space to allow for clear thinking, practical forms of assistance, and tremendous understanding. By this point, the family has likely endured countless struggles with the illness. Continuing the fight and looking towards experimental options are essential pieces of armor that families use to cope and, for some families, it may make sense to search for options as long as possible. No one can determine when a specific family should cease searching for treatment; therefore, physicians can offer invaluable support by providing information and opportunities for discussion, helping families make decisions, accepting their choices, comforting them, and remaining available.



## What Physicians Can Do

- Provide the opportunity for an initial psychosocial assessment of the child and family at the time of diagnosis.
- Refer the family to appropriate counseling and other resources throughout the life of the person with FA.
- Provide developmentally appropriate information to patients to enhance their understanding of and familiarity with FA. Encourage dialogue among children with FA, other bone marrow failure diseases, or other life-threatening illnesses.
- Encourage the family to become involved in activities sponsored by FARF. These activities aim to help families develop and maintain an up-to-date knowledge base, gain psychosocial support, and play an active role in supporting FA research.
- Help families forge working partnerships with their physicians, allowing for mutual respect for what each party brings to the situation.
- Enable patients with FA, as they mature, to become responsible and proactive with regard to their medical care.

Support after the death of a child, whether the child was very young or an adult, is essential yet surprisingly difficult to find. Rarely do bereaved parents feel that their loss is understood—and in fact, others find it difficult to understand what they are going through. Grieving parents may find it difficult to accept support, except from people who have endured similar losses. Parental grief does not go away; it changes over time. Certain factors related to the length of the illness tend to complicate the mourning process for families, such as the number of children with FA, a perceived or actual lack of support and the mourner's perception of whether the death might have been prevented. After a long, hard fight, the family may feel a sense of guilt at not having been able to prevent the child's death.

Relationships between clinicians and families should not end abruptly during the bereavement period, as it is a most difficult phase. Explaining to families that intense feelings of anger, regret, loneliness, and depression are part of the natural grieving process is often helpful. Ongoing communication may serve to reflect on the child's life, provide referrals for counseling and support groups, and express empathy about the family's struggle.

The death of a child or sibling is devastating and has lifelong implications for the family. The added complication of a genetic illness—one that a family will continue to deal with for generations to come—adds to the complexity of coping after a child dies. Fanconi anemia will always be an issue for an affected family. Many members of the FA community feel a unique connection with one another, attesting to their resilience and ability to value life and embrace the future. The FA community is strong, active, impressive, and has been greatly empowered by the FARF. Families should be referred to the FARF at the time of diagnosis so that they can avail themselves of the many services provided.

Although the diagnosis of FA can impose great challenges, it can also enable all family members to find great strength, to learn to embrace life to its fullest, and to assess what is valuable in life. The support within the FA community through FARF and its FA family meetings fosters hope in families and enables resilience on the FA journey <sup>(8)</sup>.

## Chapter Committee

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