FA family newsletter

A Semi-Annual Newsletter On Fanconi Anemia For Affected Families, Caring Physicians & Research Scientists.

NEWSLETTER #10

JULY, 1991

Fanconi Anemia Family Symposium A Success!

More than 100 FA parents and children from 24 states and 7 nations attended the first FA family symposium in Washington, D.C. on July 12–13, 1991. This represents the largest single gathering of concerned FA children, adults, parents and medical scientists in history.

Seven leading scientists gave presentations concerning many aspects of diagnosis, disease management, therapeutic trials and genetic research into FA (see following brief summaries).

Evaluations of the symposium were uniformly outstanding. Plans already are underway for a similar meeting next year. Special thanks are due to **Dr. Vicki Athens**, whose vision and persistence created this opportunity; to Coordinator **Linda Solin** whose hard work and splendid organization made this weekend possible; and to the Oregon-based **Meyer Memorial Trust**, which generously has granted funds for extensive international support efforts. (See story page 6.)

Editorial

We didn't know who'd come, or what to expect. We were overwhelmed, fulfilled and profoundly excited by what we experienced with you in Washington, D.C.

We're very diverse — by race, background, education, experience and nationality. Some — especially those who have lost children despite heroic family efforts — came with reluctance, anxiety and special feelings of personal loss.

Other families have encountered transplantation agonies with every variety of human emotion, regardless of ultimate success or failure. Many, such as we, still wait. We try to hasten the progress of science. And we manage — as best we can — the peaks and valleys of this unwelcome hateful disorder.

From the moment we entered the room we felt the instant belonging and deep bonding of everyone present. Spontaneous family group conversations, constantly growing and constantly changing continued, unscheduled, into early morning hours, long after our "official" adjournment. Discussions began anew at breakfast and through the day as — reluctantly — we said farewell.

Several casual observers asked us if this were a family reunion. It was. It was very special. This editorial message has two final purposes. The first is to thank all of you we just met personally for being our friends all these years. The second is to invite the rest of you FA personal friends whom we have not yet greeted to meet with us next time. God bless you all.

Dave and Lynn

New look for FA Family Newsletter

Notice our attractive new look! A special thanks to **Martin Sankey** and his employer, American Greetings Corporation for their donation of time, labor, material and financing to produce this edition of the FA Family Newsletter.

Editors' note and disclaimer

Statements and opinions expressed in this Newsletter are those of the authors and not necessarily those of the editors or sponsoring Fund. Information provided in this Newsletter about medications, treatments or products should not be construed as medical instruction or scientific endorsement. Always consult your physician before taking any action based on this information.

Highlights

Dear FA Families,

I am so glad to belong to the group that represents our disease, Fanconi anemia. Since Travis was diagnosed 7-1/2 years ago with FA, our family has participated in several support groups that concern children and chronic illness, yet we have never been able to discuss fully our unique problems!

I couldn't imagine that I would ever be in the same room with more than 100 people who are carriers of FA and their affected children. The first ever Fanconi's Symposium was an opportunity I will never forget. I was so interested in everyone's individual story and was able to share my own with good listeners.

One of the most gratifying parts of this past weekend was my interaction with other FA children and their siblings. What a privilege that was for me! Next year I hope my whole family will come and you will bring yours.

Marilyn Massino

From the workshop evaluations

The evaluations were uniformly positive. However, we know many will agree with one parent who wrote:

"We certainly need to hear from the doctors with regard to continued progress in all the research that's being done. However, there was nothing scheduled this year about the psychological impact on parents, child and/or family in general. There were over a hundred people here smiling and acting relatively happy while inside we are being torn apart. The impact on us as parents and on our families by this diagnosis of Fanconi's is staggering. Perhaps next year we can have some experts in the field of family counselling with a special expertise in handling serious illness and death."

Your editors agree wholeheartedly. It will be done!

Highlights of scientific presentations

I. N.T. Shahidi, M.D. Professor of Pediatrics at the University of Wisconsin

Dr. Shahidi spoke on the treatment and management of FA as a complex disorder. In addition to his review of diagnostic findings and evidence, Dr. Shahidi discussed prevention and treatment issues.

Preventive measures

1. Prevent or promptly treat viral and bacterial infections. In the case of chickenpox, Dr. Shahidi suggests treatment by zoster immunoglobulin within 72 hours of exposure. Vigorous therapy should be sought in the case of bacterial infections in order to avoid septic shock.
2. FA patients should avoid excessive exposure to sunlight. Sun blocks can be effective.
3. Dr. Shahidi suggests use of certain vitamins known to be "free radical scavengers". His recommendations are contained in a comprehensive letter he submitted to this newsletter (Appendix I). [**Editors' note:** We caution that any therapeutic recommendation always should be discussed carefully with your own physician, even in the case of over the counter vitamins or drugs.]

Disease management

Dr. Shahidi discussed at length four therapies which have or may in the future yield promising results: androgens, marrow transplantation, growth factors and superoxide dismutase.

II. Blanche Alter, M.D., Pediatric Hematologist, Mt. Sinai Hospital, New York City, N.Y.

Dr. Alter provided a helpful review of the role of medical consultants, emphasizing the primary responsibility of the treating physician.

Dr. Alter summarized major findings from the nearly 800 FA cases documented in medical literature, suggested the desirability of periodic bone marrow studies to monitor patient progress, and reviewed preliminary results of laboratory studies into the stimulation of FA cells by various "growth factors". The newly-discovered "stem cell factor" or "steel factor" showed scientifically interesting interactions with FA cells. However, therapeutic trials may be a year or two away. And Dr. Alter cautioned participants against any premature conclusions or optimism before the full process of scientific peer review has been completed.

III. Dr. Joseph Gertner, Professor of Pediatrics and Program Director of the Children's Clinical Research Center, New York Hospital

Dr. Gertner addressed issues of growth in FA children. He reviewed childhood development studies and suggested the particular importance of testing FA children for the presence and levels of naturally-occurring growth hormone.

Dr. Gertner stated that the use of artificially-cloned growth hormones might be appropriate for certain FA children: those who produce little growth hormone naturally and those who have successfully undergone bone marrow transplantation. The population most likely to respond would be those children who lack growth hormone naturally and the very young. Dr. Gertner discussed possible concerns associated with the use of artificial growth hormone. Studies are underway to determine whether this product will have safe applications for FA patients of short stature.

IV. Alfred Gillio, M.D., Pediatric Hematologist at the Sloan-Kettering Memorial Hospital, New York

IL-3 (Interleukin 3), a recently cloned growth factor, now is under intensive study in human trials at Sloan-Kettering Memorial Hospital in New York. Dr. Gillio offered promising, but very preliminary data showing a "trilineage" (red cell, white cell and platelet) growth response in at least some aplastic patients.

FA patients have been included in Dr. Gillio's first study cohort of 30. A new protocol for another thirty patients is in the planning stages. Your physician should contact Dr. Gillio to determine whether the conditions of this experimental growth factor therapy might apply to your FA child.

V. Eva Guinan, M.D., Pediatric Hematologist, Boston Children's Hospital

Dr. Guinan explored the preliminary results of GMCSF therapy for aplastic anemia. Significant results in stimulation of white cell growth have been achieved in almost all patients. Five FA patients have received GMCSF in an on-going trial, and four of the five showed a significant increase in white cell production. One patient has demonstrated a significant red cell response going from transfusion dependent to independent. Dr Guinan believes that it is premature to conclude whether this therapy

Highlights continued.

will significantly affect red cell and platelet production, although early results suggest that GMCSF affects primarily white cells in [illegible]t patients. Based on previous studies, Dr. Guinan expects that red cell and platelet response is more delayed than white cell response, as was the case in the responding patient. Remaining patients are currently being evaluated for red cell and platelet production.

VI. Lyle Sensenbrenner, M.D., Director of Bone Marrow Transplant Program, Detroit Medical Center Institute of Oncology and Allied Diseases

Marrow transplantation as a therapy for FA victims was the subject of a thorough and well received presentation by Dr. Lyle Sensenbrenner. Dr. Sensenbrenner presented a comprehensive review of transplantation technology, HLA (histocompatibility) requirements, and the special sensitivities of FA victims to marrow transplantation risks. He concluded that approximately 80% of FA patients who are prepared for transplantation using the modern approach and have HLA matched sibling donors now survive transplantation.

VII. Arleen Auerbach, Ph.D., of The Rockefeller University, New York

Dr. Auerbach concluded the formal program of scientific presentations. She noted the special importance of accurate prenatal diagnostic testing for families at risk. She further reported that studies of FA victims in the International Fanconi Anemia Registry now demonstrate that birth defects — not previously reported — can affect any system of the body in some FA victims. Most recent discoveries show that some FA children have defects in the cardiovascular system as well as the central nervous system.

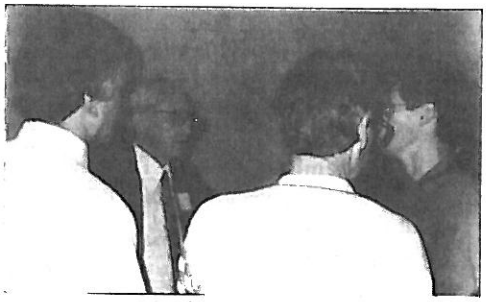
Dr. Auerbach also reported on molecular genetic research in progress. Published data now suggest that one of four genes potentially responsible for FA is located on the short arm of chromosome 20. Dr. Auerbach is now intensively studying several genes which she believes are potential "candidate" genes for FA. We expect significant reports on the gene identification progress of Dr. Auerbach and numerous other researchers supported by our Fund at the satellite conference in October, 1991.

Symposium Snapshots

◆ *Blanche Alter, M.D. discusses aplastic anemias and new research in progress.*

◆ *Arleen Auerbach, Ph.D., describes genetic research developments to intent listeners (only half of audience is pictured).*

An International Reunion, ◆
Left to right: Charles Bichet (France), Dave Frohnmayer (USA), Ralf Dietrich (Germany), Giovanni Pagano (Italy).



◆ *Dr. N.T. Shahidi (center) answers questions of (left to right) Bill Lucarell, Bob Sacks and Charles Bichet.*

◆ **Post-meeting conversations,** *Left to right: Alain Siverston (France), Janice Duffy, Margaret Curtis and the Sacks family.*

◆
Paula Ceresa, non-symptomatic FA survivor shares thoughts with families and her fiance (far right).

Medical review checklist for FA families

Suggestions for baseline medical evaluation and consultant visits for patients with FA

from: Ellis J. Neufeld M.D., Ph.D. Director, Hematology Clinic, Children's Hospital, 300 Longwood Ave. Boston, MA 02115

Many thanks to **Dr. Ellis Neufeld** for the following:

As many families are well aware, FA patients are potentially susceptible to a diverse set of medical, physical and developmental problems. Many patients have seen an army of consultants, and have accumulated a mountain of test results. This checklist is meant to serve as a reminder of important "baseline" studies, and regular consultations.

These studies and consultations are best arranged by the primary physician, who can collect the data in a centralized file, and provide copies of results and consultants' suggestions to families and to future consultants. Routine blood counts and many of the other tests may also be done by the primary pediatrician, while a few consultants may be available only at major medical centers.

Baseline tests

DEB (DiEpoxyButane)-induced chromosome breakage test. *All brothers and sisters of patients should also have DEB testing.*

Bone marrow chromosome analysis.

HLA typing. Patients, their siblings and parents should be typed at the time of diagnosis in anticipation of possible bone-marrow transplantation for aplastic anemia. Searching for *unrelated* donors need not be done at the time of baseline evaluations, but might be considered at a future time. Full blood typing for the patient should also be performed.

Blood chemistry screen. This should include studies of liver and kidney function, as well as of iron status.

Formal hearing testing.

Developmental assessment – particularly important in toddlers or early-school-age children.

Ultrasound examination of the kidneys and urinary system.

Baseline consultant visits

Genetics: Families with suspected or diagnosed FA should see a trained medical geneticist, who should:

– perform a careful, complete physical examination of the patient and any siblings,

– obtain a thorough family history, and document this in a careful pedigree,

– provide formal genetic counselling about the inheritance of FA and the availability of prenatal diagnosis by DEB testing.

Repeat genetics visits every few years are also a good idea. Physical features develop over time, so some findings of FA, such as pigment changes, may be apparent only later in childhood. Research into the genetic cause of FA is continuing, and new information about diagnosis or treatment may become available over the years.

Hematologist: Every FA patient should be closely followed by a pediatric hematologist. If blood counts are normal, visits can be relatively infrequent, but families should establish a relationship with a hematologist in any case. Most pediatric referral hospitals in major medical centers have pediatric hematology consultants, but FA is rare enough that some specialists will never have seen a case. In this event, a second opinion from more experienced hematologists is in order.

Ophthalmologist: A full examination by a pediatric ophthalmologist should be done at the time of diagnosis, with a follow-up as needed for any problems identified.

Endocrinologist: Baseline evalu[illegible] should be the rule for every patie[illegible] if hormonal abnormalities are present, or if androgen therapy for anemia is considered, follow-up evaluations are also suggested.

Other specialist visits

Patients with specific problems may also require baseline assessments and follow-up care from other disciplines:

Hand Surgeon: Serious abnormalities of the thumbs or wrists warrant referral to an expert in surgical reconstruction of the hands. In different institutions, the hand surgeon may be an orthopedist or a plastic surgeon.

Gynecologist: Adult women with FA should be seen regularly. Frequent examinations and pap smears are in order because of possibly increased incidence of gynecologic malignancies.

Urologist/Nephrologist: For patients with significant abnormalities of the urinary system.

Notes:

1. This list is not all-inclusive. Each specialist will suggest further b[illegible] tests, X-rays, or other evaluation[illegible]
2. The suggested tests and visits are based on problems in FA which have been reported in the medical literature. It is important to remember that medical journal reports of rare diseases like FA may have an "ascertainment bias"—that is, a tendency to report cases more severe than average. Thus, a majority of FA patients will have mostly normal baseline studies; obtaining these tests is important nevertheless.

Treatment of Fanconi's Anemia

The Clinical Hematology Branch of the National Heart, Lung and Blood Institute is conducting research in Fanconi's anemia. Patients will be admitted for research studies and will then receive enzyme therapy consisting of superoxide dismutase by the intravenous route for two weeks. The premise of this treatment is to correct a defect in radical scavenging, failure of which results in DNA damage. The toxicity is anticipated to be absent or low. If you have patients with Fanconi's anemia who would be willing to participate in such a study, please contact either **Dr. Johnson Liu** or **Dr. Neal Young** at the following address:

Clinical Hematology Branch
National Heart, Lung and
Blood Institute
Building 10, Room 7C-103
9000 Rockville Pike
Bethesda, Maryland 20892
(301) 496-5093

Protocol allows for 10 patients.

Anxiety does not empty tomorrow of its sorrow, but is does rob today of its strength.

Anonymous

FA Families Continue Fund Raising Efforts

Many FA families and their friends have worked hard to generate badly needed funds to support medical and genetic research. We are deeply grateful for these wonderful efforts. This newsletter will report only on funds raised from January through June, 1991. Many families raised substantial sums which have been reported in previous newsletters.

Phyllis Cafaro continues to amaze and impress us with her persistent efforts and stunning results. During the past six months she has raised $44,938.15 to support research. We are all deeply indebted to Phyllis for her dedication and determination to impact this difficult illness.

Deane Marchbein and **Stuart Cohen** raised $26,161; **Vicki** and **Andrew Athens** generated $21,775 and **Ed** and **Barb Brookover** raised $20,725. All of these efforts represent countless hours of hard work and the generosity of caring friends and relatives.

Dr. Sidney Farkas and his wife, **Ethel** raised $11,531; **Ron** and **Fredi Norris**, $9,670; **Marlene Stone**, $9,489.84; **Sandy** and **Marc Weiner**, $9,012; **Pat** and **Bill Danks**, $7,580; **Richard** and **Dotty Day**, $7.831.50; **Ed** and **Janice Duffy**, $4,675; **Elaine** and **Kevin O'Connor**, $1,800; **Bill** and **Jackie Lucarell**, $1,400; **Lynn** and **Dave Frohnmayer**, $1,548; **Alan** and **Kathy Bresette**, $1,250; **Leardon Keleher**, $1,000; **David and Christine Westmoreland**, $500; **Akhtar Mansoor**, $500; **Ceresa family**, $870; **Wilfredo** and **Carmen Gonzales**, $200; **Robert** and **Linda Scullin**, $440; **Irene** and **John Kalman**, $500; **Nora Herren**, $110; **Sandy** and **Eddie Allen**, $183.73. Other FA families made individual contributions which are deeply appreciated as well.

Each family in the support group has a different capacity to generate funds for research. Please know that each and every effort is invaluable, and will help us to understand and conquer this devastating illness. Please continue with your wonderful efforts!

On a sad note, funds were also contributed to the FA Research Fund as a memorial to a child or young adult who passed away from complications of FA. Funds were contributed on behalf of **Avi Weiner**, **Dennis Oster**, **Dee Dee Doutt**, **Brian Logsdon**, and **Caleb Morgan**. We are deeply saddened by these tragic losses.

Dear FA Families and Friends:

I know you share our commitment to finding a cure for this devastating disease of children. Thanks to your fundraising efforts, Fanconi Anemia Research Fund, Inc. currently supports nine crucial research projects. But the need continues. We want to eradicate this disease in order to spare the precious lives of our children. The Board and staff of Fanconi Anemia Research Fund, Inc. are committed to raising research dollars which we hope will ultimately provide the bright future our children deserve. Here's how you can help:

1. Part of our public awareness strategy is to have a national spokesperson. Some of you may have contacts with celebrities or other well known people. If you know of someone who may be willing to be a celebrity spokesperson, please contact me or **Linda Solin**.
2. Please continue with your fundraising efforts. If you have not raised money for research, please consider doing so. I have designed a fundraising brochure for your use. This brochure describes our organization, the disease, and explains the need for research funds. If you would like a copy, please contact Linda Solin. Let us know how many you can use.
3. Soon I will have a packet of fundraising ideas available for you. I hope this will inspire you to begin raising much needed research dollars. Linda Solin can also provide you with sample fundraising letters for your use.

I can be reached at the following address:

Phyllis Cafaro
c/o Cafaro Co.
2445 Belmont Avenue
P.O. Box 2186
Youngstown, OH 44504

Please feel free to contact me or Linda Solin with your fundraising questions, ideas, or needs.

Sincerely,
Phyllis Cafaro
Board of Directors
Fanconi Anemia Research Fund, Inc.

Tiger Charities Challenge Met

As many of you know, last February, Fanconi Anemia Research Fund, Inc. received a two part research grant from Tiger Charities. The Fund was awarded $60,000 for gene identification research. In addition, we were awarded a "challenge grant" in the amount of $60,000. This "challenge grant" was to be awarded to us on the condition that we first raise a matching $60,000 for our Gene Identification Project.

Many of you raised funds for this effort. We are thrilled to announce that we have met the $60,000 challenge. Soon we will have more than $180,000 to be used exclusively for gene identification. This comes very close to our annual need of $246,000 for genetic research. Thank you!

We especially thank the **Norris family** for their role in helping us secure this grant. Gerald Norris works for Tiger Management Corporation, the parent company of Tiger Charities. The Norris family contacted us about Tiger Charities, made the suggestions about the grant proposal, and facilitated the application. **Dr. Blanche Alter** was also instrumental in securing this grant for us. She left her laboratory on very short notice to deliver lucid and persuasive testimony on the importance of genetic research.

If you work for companies which offer charitable contributions, please let us know! We can write grants and provide any documentation needed, **but we need your guidance in finding these organizations.** We desperately need to apply for treatment funds to support the many investigational treatments which are now opening up. Please let us know if you can help us to raise these funds.

FA Research Fund, Inc. Receives Administrative Grant

The Meyer Memorial Trust recently awarded a three year grant totalling $197,000 for the FA Family Support Program. This award will fund the newsletter and most office overhead for three years. In addition, it will assist us in achieving the following objectives:

1. Broaden recognition and early diagnosis of this disease, as well as increase the identification of previously diagnosed but unserved FA victims

Fanconi anemia occurs in all ethnic populations and all geographical areas, yet our support group population reflects that California and other major population areas are underserved. We will develop comprehensive awareness including selected visits in approximately five major medical centers or teaching hospitals per year.

Our two scientific workshops reveal consensus that Fanconi anemia is underdiagnosed because practitioners are unfamiliar with its now obvious manifestations. We will facilitate or encourage the publication of three journal articles per year for general practitioners or practitioners in relevant medical specialties. This strategy will speed diagnosis by internists, plastic surgeons, pediatricians and others who should recognize the disorder before bone marrow failure is ultimately detected.

2. Broaden treatment options for FA families

The FA Support Group has already served as a primary resource for the testing of exciting new drugs and therapies that have lifesaving potential for FA victims. We will continue to serve as a resource for experimental drug and research protocols.

3. Provide traditional family support

We will continue and strive to increase our family support outreach by a minimum of 10% per year. **Geri Young**, our intern from the University of Oregon, has completed her degree and has been hired for the grant-funded Family Support Coordinator position.

The Meyer grant will also enable us to offer a Family Support Symposium on a yearly basis. We will continue to offer the newsletter twice annually.

Upcoming conferences

Pediatric hematologists to hear Fanconi anemia presentations

Thanks to the efforts of **Dr. Michael Greenberg** and his Pennsylvania-based Fanconi Anemia Research Foundation, the nation's pediatric hematologists will hear a major presentation on FA.

The American Society of Pediatric Hematology will be given a half-day symposium on FA and its manifestations at a meeting in Chicago, Illinois on September 12–14, 1991. We salute Dr. Greenberg for arranging this important outreach effort, which representatives of our Fund will attend as well.

FA gene researchers to meet

Approximately thirty scientists from around the world will attend a "satellite conference" on Fanconi anemia genetic research progress in Washington, D.C. on October 5–6, 1991.

Dr. Arleen Auerbach, Ph.D., The Rockefeller University, and **Dr. Manuel Buchwald, Ph.D.**, of the Hospital for Sick Children, Toronto, Canada are co-organizers of the satellite conference, to be held just prior to the 8th International Congress of Human Genetics. The Fanconi Anemia Research Fund, Inc. agreed to provide financial support to make the satellite meeting possible. Our representatives will meet with the scientists to reinforce the urgency of our common objectives in understanding this disorder.

Because of the significant numbers and quality of scientists who will attend this conference, the Fanconi Anemia Research Fund will not need to sponsor a separate annual scientific workshop this autumn.

Orphan Disease Conference

The 1991 NORD Patient Family Conference will take place in Baltimore on August 16 and 17, 1991. The National Organization for Rare Disorders (NORD) has planned a quality meeting for families and patients affected with rare diseases. Members of the FA Family Support Group qualify for a discounted registration fee. (The FA Research Fund is an Associate Member of NORD). If you would like more information about the conference, please contact **Geri Young** at this office (503) 687-4658. She can send you a registration form and a schedule of events.

Kids' Corner;

Send your articles!

Your editors were asked at the Symposium to set aside a section of future newsletter issues for a "Kids' Corner". If your FA children or their siblings have articles, questions or ideas, we will publish them, space permitting. Please encourage your child to participate!

Free medically-related travel

If you are flying to another city with your child for his/her medical treatment, please let us know. We have been successful in securing complimentary flights for FA patients and their parents. We would like to assist you, should you have this need. Contact **Linda Solin**, (503) 687-4658, at this office.

Make A Wish Foundation trips

Have you considered referring your child to the MAKE A WISH FOUNDATION? Most FA children do meet the criteria of this and other similar wish granting foundations. If you would like to have your child referred, feel free to call **Geri Young** or **Linda Solin** at the Fanconi Anemia Research Fund, Inc. office (503) 687-4658.

Autosomal Recessive Genetic Disease:

An explanation

Many families know that Fanconi anemia is an "autosomal recessive genetic disease". But what is that?

The explanatory article in Appendix II is included for three reasons. First, no spouse should "blame" the other, or feel personal guilt. All persons carry several, and perhaps many lethal genes. Only when two carriers mate does the chance of an affected child even occur. Second, FA carriers presently have no way of knowing in advance of their condition. It may not have appeared in your family line for dozens of generations. And finally, understanding how the disease is carried has important implications for family planning. These should be discussed with your family doctor or genetic counselor.

The article is excerpted, with our thanks, from the Spring 1991 issue of *TEXGENE*.

Q&A

Diana Fitch is a child therapist who has worked with post-trauma children. She is the mother of three sons, one of whom has FA. Diana has agreed to write a regular column for our newsletter concerning the psychological aspects of living with FA. She invites you to send your questions or concerns to the FA Research Fund and she will respond in our next edition. Many thanks, Diana, for adding this crucial dimension to our newsletter!

About Kids — The non-medical side

Q. Since our baby has been diagnosed with FA our older child seems different — unhappy and distant. What can we do?

A. You're on the right track by noticing that your non-FA child is also affected by this disease. By accepting your child's feelings as normal and acknowledging them within your family, those emotions that previously have not been discussed will become less confusing and less powerful. Feelings of guilt, anger, resentment and even jealousy are all possible. Listening to your child's particular concerns and fears — without passing judgement on them — will help you know what actions to take to reassure him.

Every child has wished harm to his sibling. The FA diagnosis can make those wishes seem like they are coming true. One little boy stated he had caused his sister's illness because several years before she had broken her arm while they played together. This broken bone in his mind was intertwined with bone marrow failure. Children must hear clearly that FA is no one's fault — not the FA child's, not the siblings', not the parents', not anyone's.

Your healthy child's identity can sometimes seem eclipsed by his status as the sibling of an FA child. Help maintain your child's interest (and your interest) in his hobbies and activities. Allow your child's friendships outside the home to continue even during crisis periods.

Make special time for you and your child to be together. For the young child it might be ten minutes each evening to review the day. For the older child it might be a monthly luncheon. One school-age child stated his favorite time was to help with the weekly marketing and then stop for a soda on the way home. Do this consistently so your child can rely on getting your attention positively. If your child's suffering continues despite your efforts to relieve it, consider consulting professional psychological help.

Diana Fitch, Child Therapist

From our families

Pamela Baxter (314-491-3551) writes of the shocking discovery last September that sons **George** (10) and **Bradley** (6) have FA. Another son, **Stuart**, happily was a perfect HLA match for George. George's transplant at CHMC in Cincinnati occurred in late January and, as of this date, is progressing well. Pamela writes:

"George entered CHMC on Jan. 21 for his transplant. January 30th Stuart's marrow was harvested and given to George. Because the risk of infection is so high, George was placed in isolation until his blood showed signs of engraftment which took only 10 days (a new record for FA at CHMC). We were home February 23rd for supper with the entire family. George is still on drugs and seen by doctors. It will be awhile before he will be off all drugs and normal, but I feel the hard part is over, thanks to the staff of CHMC."

The Baxters anticipate that son Bradley will need an unrelated marrow transplant. While acknowledging the higher risk involved, Pamela stated that she had "no reservations about going ahead with another transplant after watching the staff work to make George's transplant a success."

Jan and **Richard Turner** of Auckland, New Zealand wrote us recently about their wonderful daughter, **Kelly May**. Kelly was born with esophageal atresia (esophagus disconnected from the stomach), tracheal malacia (softening of the trachea), two thumbs on her left hand and no tendon in the thumb on her right hand. She was four weeks premature and weighed 3 lbs, 7 oz.

The Turners wrote at great length of the many surgical procedures Kelly endured in order to correct successfully her various anomalies. When Kelly was six, she was diagnosed with Fanconi anemia. The Turners are hopeful that a bone marrow transplant under **Professor Elianne Gluckman** in Paris, France will correct her present bone marrow failure.

The Turners write *"Kelly is now 8 years of age and is in Standard 2 at Taupo Primary School. She is a happy, lively and loveable little girl who, although only the size of a 5 year old, lives life as fully as any other child of her age. Her platelet count is only a fifth of the normal count but she is keeping good health at present. For how long — we don't know."*

Dear FA Friends:

Last February our seven and a half year old daughter **Alaina** had the privilege of going to Los Angeles and meeting the actors on FULL HOUSE. It was an exclusive arrangement in that **Stephanie** presented Alaina with a Michelle doll. We watched them do some filming and talked to them between the shootings. We did this for about two hours. The next three days we were treated to Disneyland and Universal Studios. The reason we are sharing this with you is that the Starlight Foundation grants wishes to children who have a "life threatening" disease. Most of us FA families more than qualify in that respect!

From left to right: Mary Kate Olson, Candace Cameron, Alaina Riley, Jody Sweetin, Leonard Riley III.

Starlight was founded in the early 1980's and has granted over 4300 wishes. They are a first class group of people. Every year they hold balls and galas where actors (stars) can attend. From these events they derive most of their contributions. If you would like to contact them for further information you can call 1-800-274-7827. You may want to mention that you heard that they were the best wish granting foundation and that you wanted to make an inquiry. We realize there are other qualified organizations that grant wishes to children. And they do a fine job. However, if you or your family have not been provided with a telephone number of one, and would like to pursue a wish, we can personally recommend Starlight.

Alaina will enter the hospital on Wednesday, July 31 for her transplant. We attempted to do this in May, but she developed some complications of the liver. At the present she is stable enough to proceed. **Lenny,** her five year old brother is the donor. We will be following the Gluckman regimen. We ask that you keep her in your thoughts and prayers. Should

Continued on page 8.

you like to send her a card the address is:

Cook's Children's Hospital
BMT Unit/Alaina Riley
801 Seventh Avenue
Ft. Worth, TX 76014

I would like to take this opportunity to thank **Linda Solin, Dr. Vicki Athens** and the others who put together the FA Symposium in Washington, D.C. Jan and I were thrilled to meet so many of you. Whether we like it or not we are tied together by a damaged genetic chain. It was comforting to see the level of strength that many of you displayed as you fight your own individual battles. The information from lectures is already proving to be invaluable. Thank you to all.

We will conquer FA,

Leonard, Jan, Alaina, and Lenny

Memorial Observations

Our hearts go out to **Moira** and **Charles MacLellan**. Their sons **John**, and now **Danny** have been claimed by FA. Danny passed away on December 26, 1990 from post-transplant complications.

Moira wrote touching comments about her boys. But beyond that, she offers thoughtful and important reflections on parents' relationships with medical institutions and providers. We thank Moira for her insights:

"Danny was a complex little fellow, with a very honest way of looking at life. He loved wrestling, yet he also wanted to make things and enjoyed knitting for the past year or so. He worried about the possibility of war and about social problems. He had a lot of "guts", and at times, I was a little taken aback by his honesty. I don't believe he ever lied in his life.

During the many uncomfortable procedures he often had to endure, he never shed a tear although he often became somewhat grumpy. He would also demand to know all details before reluctantly agreeing to necessary medical procedures.

Charlie and I have learned so much from the short lives and tragic deaths of both our boys. John, Danny's older brother died in March 1989 at the age of nine from a brain hemorrhage. He was also a victim of Fanconi's. Danny was only seven. The honesty, tolerance and love shown by our children was a lesson to us, their sisters and our friends and family. Many who knew them remarked on their bravery in the face of such severe illness and pain.

How we all wish that things could have been different and that they were still with us. Still, although their loss hurts beyond words, I'm so glad we had the chance to know and love them even for their few short years.

From a practical point of view and drawing from our experiences over what seems like a lifetime, I have some advice which may be of help to other FA families:

1. *It is most important to establish a mutually satisfying and open relationship with the doctor(s) primarily responsible for your child's overall care and well-being. If this is not possible find another doctor and if that's not possible, be extremely careful that personalities are not getting in the way of treatment.*
2. *Make sure that one person is responsible for primary care and don't get moved from one person to another so that no one really knows what is going on.*
3. *Recognize that you are your child's best advocate and have confidence in your ability to fulfill the demands of that role. Never lose sight of the fact that you know more about your child and his or her individual needs. If you feel these needs are not being properly attended to, demand the necessary attention or treatment.*
4. *One of the most essential components, if not the most essential component, in obtaining satisfactory care is to establish an open channel of communication between you and all parties involved in your child's care. This is crucial especially before the commencement of any significant or critical medical intervention, and becomes even more important where more than one medical facility or unit is involved.*

Be ever vigilant looking out for your child's best interests. As Bone Marrow Transplants are, by their very nature, at the leading edge of medical technology and science and, as each patient has his own unique response to treatment, communication between parents, family doctors and specialists is most important to the life and health of your child. At the risk of repeating myself, I urge you never lose sight of this fact."

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Appendix I

As you are well aware patients with Fanconi anemia are at significant risk to develop leukemia and other forms of cancer. The reason(s) for the high incidence of malignancies in Fanconi anemia patients is (are) not clear. Increased chromosomal breakage and hypersensitivity to toxic oxygen species known as superoxides and free radicals may be responsible. Indeed the type of leukemia encountered in FA patients is almost entirely of myeloid origin and similar to that seen following radiation or administration of chemotherapeutic agents. These agents are known to damage the genetic material in cells (DNA) by producing reactive oxygen molecules. Environmental carcinogens such as passive inhalation of cigarette smoke, excessive exposure to ultraviolet light and ingestion of meat containing high concentrations of sodium nitrate may also play a role.

Numerous studies have shown an increased relationship between the risk of cancer and the consumption of beta carotene (vitamin A precursor). Lower intake or lower plasma concentration of beta carotene have been found in persons who subsequently developed cancer. For instance, a number of epidemiological studies indicate significantly increased risks of cervical pre-malignant or malignant lesions in women with low blood levels of beta carotene. On the other hand, it has been shown that the administration of beta carotene or related retinoids can prevent various types of cancer. In a most recent, and rather convincing report, it was demonstrated that a vitamin A derivative, isotretinoin, clearly reduced the incidence of secondary malignancy in a large number of patients who had received radiotherapy and/or chemotherapy for head and neck cancer. It is of interest that in tobacco chewers, there is a chromosomal instability which is however restricted to the mouth. It has been found that beta carotene or vitamin A prevent or result in complete remission of the precancerous lesions of the oral cavity in these individuals.

Separate investigations have provided evidence that vitamin C and E containing foods are also associated with lower risks of certain types of cancer. For more information on the protective role of the above antioxidant vitamins against cancer, I have included a list of the most recent pertinent publications in this area.*

While isotretinoin (13-cis-retinoic acid) has some side effects, such as skin dryness, eye inflammation, and increased blood fat levels, beta carotene, vitamin E, and vitamin C are devoid of toxicity at the dosage commonly used to prevent cancer. Although it has been reported that vitamin E at the dosage of 400 U per day may result in decreased platelet adhesiveness, clinical bleeding manifestations have not been reported in individuals receiving even very high dosage such as 600-1000 U per day. Since the potential benefical effects of the vitamin intake in patients with Fanconi anemia by far outweigh the possible side effects, randomized studies using placebos seems, for ethical reasons, unwarranted. The incidence of cancer in FA patients within the next decade could be compared to the historical controls and information available through the FA registry. In addition to the possible role in cancer prevention, the above vitamins may exert a beneficial effect on growth and development and possibly hematologic status in the patients. It is well known that in the body, three organs namely hemopoietic (bone marrow), male gonads (testes), and the gastrointestinal tract have the highest rate of cell turnover (increased rate of DNA synthesis). As a result, these organs are extremely vulnerable to agents which produce reactive oxygen molecules, such as radiation. Although bone marrow failure and hypogonadism are well known complications of FA, the functional abnormalities of gastrointestinal tract have not been investigated in FA patients, and whether they absorb these vitamins adequately remains unknown. Consequently, baseline levels of these vitamins and periodic measurements of their levels after daily supplementation is highly desirable.

After careful investigation, we found that Carlson Laboratories, Inc., Arlington Heights, IL, well-known for the quality of their vitamin preparations, markets a combination product containing all the three antioxidant vitamins in addition to selenium. This latter trace metal is part of glutathione peroxidase, which is a well-known antioxidant enzyme. Each soft gel contains 5000 units of beta carotene, 500 mg of vitamin C, 200 IU of vitamin E, and 100 ug of selenomethionine. According to the available data, 2 soft gels should provide the necessary antioxidant activity in adults and older children. The children under 10 years of age should receive one soft gel. For younger children who cannot swallow the soft gel, the tip of the soft gel could be snipped with a pair of scissors and the content should be squeezed onto a teaspoon containing some of the child's favorite food (ice cream?). For children less than 2 years of age, the content of 1/2 soft gel capsule could be administered. Above dosages may be adjusted on an individual basis according to the blood levels.

Since the determination of the concentration of these vitamins in the blood requires sophisticated methodology and experience, we selected MedPath Laboratories, Inc., which has branches throughout the country, and is well-known for the reproducibility and reliability of all its biochemical tests. Fortunately, this company has agreed to measure the blood levels of these vitamins in FA patients at a reduced cost, which will be, in all likelihood, covered by your insurance. It is anticipated that each patient will have a baseline level prior to initiation of vitamin therapy. A second determination will be done one month after therapy to ascertain adequate absorption. Subsequent determinations will be performed biannually. For those patients who have updated growth and development charts, growth and development will be recorded at least every three months on percentile curves. This will enable us to compare the velocity of linear growth and weight gain before and after vitamin therapy.

I urge you to share the above information with your physician. Should you and your physician decide to start your child (or children) on this combination vitamin therapy, please contact **Lynda Mea** (608/263-6200) or myself (608/263-6202) so we can send you the necessary forms.

Sincerly,

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Appendix II

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Basic Information on Autosomal Recessive Inheritance

by Sandra Grilliot, M.S.

How hard is it to answer a parent's bewildered cry of "but there's no history of it on either side of our families" when a child is diagnosed with a genetic disorder. The sporadic inheritance of most chromosomal anomalies and multifactorial conditions is quickly understood, if not liked, by most parents. But the "hidden gene" concept behind autosomal recessive conditions is less clear to the public. Frequently, parents learn the autosomal recessive risk that affects their family, but are not aware of how this risk was derived. Here then is a brief primer of autosomal recessive inheritance.

To begin with the basics, the genes are the individual functional units of heredity. Each chromosome is made up of thousands of genes. The two can be thought of together, the genes like beads on a string which form a chromosome. Chromosomes come in pairs, one member of each pair coming from each parent. The two chromosomes in the pair look very much alike and are formed so that for every gene on one member of the chromosome pair, there is a gene that does the same job on the other chromosome of that pair. Therfore, we have two genes that function together to provide each bit of genetic information. While each of the two genes provides information for the same job, they provide it in different ways. This is the subtle variation that keeps us all a little different from others even though each of our bodies works in basically the same way.

It is estimated that 50,000–100,000 genes make up the 46 chromosomes. Biostatisticians have calculated that among these 100,000 working genes, everyone has 5–8 genes which are "non-working", that is, they do not properly relay the information on how to do their job. Fortunately, since we have inherited two genes for almost every job, if one does not work correctly, the other one can often compensate and still get the job done. We call a person who has one working and one non-working gene making up any particular pair a "carrier" for that non-working gene. Because this person's working gene compensates and gets the job done, this person has no health problems related to the non-working gene. But a carrier does need to be aware of their status because of the implications it could have for their offspring.

If two carriers decide to have a child together, they could each pass on their copy of the non-working gene to the child. This child then does not have a working gene to compensate for the non-working one. Therefore the body has no working gene to do whatever job that gene pair controlled. That job then is not performed properly and a disease state results in the child. Genetic diseases inherited in this autosomal recessive fashion include Tay-Sachs disease, sickle cell anemia, PKU, and cystic fibrosis.

For a couple who each carry a recessive gene, there is a 25 percent (one in four) chance with each pregnancy that the child would inherit the recessive gene from each parent and have the disease. There is also then a 75 percent (three in four) chance with each pregnancy that the child would inherit at least one working gene and so not have the disease.

Autosomal recessive diseases can only occur if both parents carry the same non-working gene. If one parent is a carrier for a non-working gene, but the other parent carries two working genes for the same job, then their offspring are not at risk for the disease since they will always inherit at least one working gene.

The availability of prenatal diagnosis for a carrier couple depends on the disorder involved. Some conditions are not prenatally detectable at this time. Others may be detected either by looking directly for the gene (as in DNA testing for cystic fibrosis) or by looking to see if the gene's job is being done properly by looking for the gene's products (as in biochemical analysis to see if babies at risk for Tay-Sachs disease are making enough Hex A).

There are other prenatal counselling points couples should be alerted to. First, certain disorders may be best looked for by certain techniques at certain stages of pregnancy and this may determine what type of testing is most suited to their needs. For instance, if a condition is best looked for by biochemical analysis of amniotic fluid obtained at 16 weeks gestation than CVS at 10 weeks gestation may not be the test of choice. Second, prenatal diagnosis is usually only offered for the diseases a couple is known to be at increased risk for. Most labs do not routinely run tests for specific recessive disorders unless notified in advance that both parents are documented carries of the non-working gene and are requesting the testing to be done. Therefore, no series of prenatal tests can look for all potential genetic disorders. Finally, the most important point, of course, is that the decision to or not to pursue prenatal diagnosis must be made by each individual couple based upon their own values and beliefs.

Appendix III

Kirsten Frohnmayer's graduation speech, June, 1991

Kirsten Frohnmayer, Student Body President of South Eugene High School, delivered remarks at her graduation ceremonies on June 8, 1991. In response to numerous kind requests from Family Symposium participants, we reprint Kirsten's speech from the video which was shown at our opening dinner meeting in Washington, D.C.

Reflections

Good evening and happy graduation! Over the last few months, all of us in the class of 1991 have taken a hard look at who we are and what we want from our lives. We have tried to match that personal inventory with a plan for the future. Because of unique circumstances, unfortunately not ones I would have chosen for myself, I believe I have gained a different perspective on how to evaluate life and how I want to live mine.

My dad confided to me recently that at one point, he wasn't sure he would ever experience the joy of seeing his first child graduate. This is because I have a potentially fatal genetic illness called Fanconi anemia. This condition eventually causes bone marrow failure, leaving one vulnerable to life-threatening bleeding, fatigue, and infection. In other words, of all the glitches that could occur genetically, this is one you might want to avoid.

I was diagnosed with this illness at the age of ten. My twelve-year old sister was diagnosed soon afterwards with the same illness. During these past eight years, we have been through a lot. I'd like to share with you tonight some of what I have learned in the hope that my reflections might prove valuable to you.

I've learned to appreciate what I've got. When all else is going badly, I've got my family and my friends for support. I'd like to encourage you to appreciate what you have. The song line *"you don't know what you've got 'til it's gone"* is really true. Too often we take for granted what is going well and focus on the negative. In the process, we lose sight of the best aspects of our lives. I'd especially urge you to appreciate your health if you've got it. So many people don't realize the importance of good health, and they throw it away. You are lucky if control of your body, your life, your future, is not dictated by your genetic makeup. It has always amazed me, having to fight for my life at times and having watched my sister fight for hers, that other, healthy people don't even think twice

FA family newsletter

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Appendix III; Kirsten Frohnmayer Continued.

before they drink and drive, fill their lungs with nicotine, or engage in other reckless behavior. You are so lucky to have a healthy body. I hope you won't pollute it. I hope you won't take chances with your life. I hope you won't take your life for granted. It is precious, and can easily be taken away from you.

I've also learned the importance of having an optimistic outlook on life. Being depressed is really not fun, and it certainly doesn't get you anywhere. All of us face adversity as some point in our lives. Dwelling on the negative, however, though seductive, is an exit into the all-consuming trap of self-pity. Focusing on more positive aspects of your life and on the pluses of a seemingly bad situation will let you enjoy life more. A way to focus on the positive is to refuse to be a victim; in other words, to take an active stance. Although it is normal to grieve and feel anger in a bad situation, working to solve your problem is ultimately more productive and satisfying. My family, for example, has formed a support group and a research fund. Talking to doctors about experimental drugs and new breakthroughs has really been therapeutic.

I have learned to keep my problems in perspective. There's always something worse that could have happened but didn't. My family jokes that by having this serious health problem, we provide an important community service. We remind people that things in their own lives may not be as bad as they seem. This probably strikes a chord with some of you parents. I bet you've said at times *"how could I have raised such a slob?"*, *"how could he think Def Leppard is better music than Mozart?"*, or *"her grades really should have been better"*. Those may be legitimate concerns, but in the scheme of things, they're not that serious. Try to live by the old saying *"Don't sweat the small stuff"*, and the truth is, most stuff is small stuff. Parents, see the positive in your kids. Fellow classmates, see the positive in yourselves.

A final thought I'd like to share with you tonight is my belief that sometimes we should live for the day. Too often life consists of anticipation of the future or regrets about the past. But we can't change the past, and we don't know what the future will hold. So, at least some of the time, we should concentrate on the present. Whatever path you've chosen, whether you're talking about college, a job, volunteer work, or family, you're talking about life, and life must be fun. Find the fun in life, for **Ferris Bueller** said on his day off, *"life moves pretty fast, and if you don't stop and look around once in a while, you are going to miss it."*

So, as you complete that self inventory, I hope that you will remember to appreciate and protect what you have, be optimistic and constructive in the face of adversity, and stop to smell the roses. Good night and good luck!

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