

THE WHITE HOUSE
WASHINGTON

October 17, 1996

Patricia Steinitz
314 Richey Avenue
Pittsburgh, Pennsylvania 15214

Dear Patricia:

Thank you for taking the time to write the President and for sharing the letter on SSI with me as well.

It is very important for us to hear from individuals like yourself who have valuable information to contribute.

Again, thank you for writing.

Sincerely,



Carol H. Rasco
Assistant to the President
for Domestic Policy

*from H-100's
or
FE-416.926 AC7*

I would like to discuss what you can do to help us. I would also like to discuss what the Social Security Administration can do to insure that children like Hunter do not fall through the cracks. I have been told by various advocates that the current childhood "medical impairment list" used by SSA does not include many rare disorders and that it does not even include a listing for skin disorders for children! I am not aware of a type of listing for life threatening disorders, either. I am also concerned that a new and more strict definition of functional abilities or age appropriate behaviors will eliminate children like Hunter.

While Hunter is doing quite well in her development, she is always at risk to falling behind quickly and drastically. At five months of age she could not lift her head up to look around. Hunter will be two years old this month and has learned to crawl, walk, use her hands within limits, and is learning to talk. We, along with the help of Early Intervention services, have worked long and hard to teach Hunter the skills she needed to accomplish these goals.

We still have a long road ahead of us since her disorder is lifelong. Hunter spent two days in the hospital last week having a G-tube (Percutaneous Endoscopic Gastrostomy tube) inserted in her belly so we can assist her consumption of the extremely high caloric and protein diet that her body needs. A combination of not feeling well and typical toddlerish behavior resulted in a weight loss of 13 ounces in the one and one half weeks preceding this procedure.

We will have to teach Hunter to eat and she will need help with her speech since she cannot close her lips due to her skin disorder. While Hunter is not affected mentally, she will have a lifelong battle with numerous problems to include nutrition, growth, infection, heat tolerance, hair growth, and motor skills. She will have to be meticulous with her skin care to prevent problems with eyesight, hearing, and life threatening infections. This is a very debilitating illness. Yet Hunter could lose her medicaid coverage because her disorder is not on the SSA "medical impairment list" and her impairments might not be considered severe enough under the new definition of disability.

~~Dear Mr. President:~~
Dear Patricia:
X you for taking time
to write the Pres. & for
sharing ~~the letter~~
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me as well. It is very important for us to hear from
individuals like yourself
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cc of response
w/ this orig
packet to alana
Patricia Steinitz
314 Richey Avenue
Pittsburgh, PA 15214
(412)-231-1201
(412)-231-3159 Fax

September 30, 1996

The Honorable William J. Clinton, President
The White House
Washington, DC 20500

Dear Mr. President,

Again, thank you for writing.

Sincerely,

I am the mother of the baby girl whose picture you saw while shaking hands with the audience at the end of your speech at Robert Morris College in Pittsburgh on September 25, 1996. My name is Patti Steinitz and my daughter's name is Hunter. It was a pleasure to meet you that day. Thank you for stopping to look at Hunter's picture. I also want to thank you for sending your aide, Stephen Goodin, over to speak to me.

CHH
File

Although I wrote to you previously and received a response from the White House, I feel it is extremely important to communicate with you directly regarding the changes to the Supplemental Security Income program for disabled children brought about by the elimination of the Individual Functional Assessment test through the Welfare Reform Bill. This is the test that qualified Hunter as eligible for SSI.

Since you, ultimately, will approve the Social Security Administration's new disability test and definition, I would like to meet with you as soon as possible to discuss the ramifications of the SSI changes that will affect not only us, but hundreds of thousands of children in the USA. It would be most beneficial for you to visit our home for a day so that you can see, first hand, what we deal with on a day to day basis. The life of raising a disabled child is extremely stressful physically, emotionally, and financially. While Hunter's disorder is unique, I find it hard to believe that we are the only family in the USA whose child's life could be endangered and who could be forced onto Welfare as a result of this bill.

I have enclosed copies of my original letter and the response from Carol Rasco, a recent Washington Post editorial, two newspaper articles about Hunter, articles from medical journals about her condition, and photos of her. You will recognize the framed part of the photo of Hunter in her ball cap which you saw on September 25th. Contained in this mailing is the complete photo which shows one of her birth photos in front of her. The items included explain background information about Hunter's disorder, how the new law will affect us, and give some insight into what we, as a family, are going through. However, no one can know the reality of the situation unless they walk in our shoes.

Before Hunter's birth, my husband and I were a normal working couple raising an 11 year old daughter and anticipating the birth of our new baby. We are both college graduates and work full time. I graduated from Indiana University of Pennsylvania and have worked as a police officer for the past 16 years. My husband, Mark, graduated from the University of Pittsburgh and is a businessman struggling at starting a new business. Hunter's birth, with her extremely rare disorder and the accompanying medical problems, has turned our lives upside down and thrown us into a new and different world; a world where our daughter's life may be threatened and we may be forced onto Welfare. It is ironic that our situation seems to defeat the purpose of the Welfare Reform Bill.

Hunter is a happy, sweet, and loving child. We love her dearly. She is an integral part of our family and of our large extended family. I will do my best to insure that nothing interferes with her medical care. I will also do my best to insure that my husband and I will be able to provide for her. It is incredible that we should have to fight our government to keep our jobs and Hunter's medical coverage, in order to accomplish these goals.

As you can see, I have become a very active advocate on my daughter's behalf. Even so, the publicity stirred through the newspaper articles and television coverage, and all the letters I wrote to the Senators has not done much good. No one appears to be listening. So now I am turning to you for help. I truly believe that you are committed to helping children and families. Will you help us?

Thank you for the opportunity to communicate directly with you. I look forward to your response and meeting you.

Sincerely,

Patti Steinitz
Patti Steinitz

cc: WTAE TV, Pittsburgh
Ellen Perlmutter, Pittsburgh Post-Gazette
Jocelyn Stewart, Los Angeles Times
Stephen Goodin, Aide to the President
Carol Rasco, Director of Domestic Policy Council
Franklin Raines, Director of OMB
Shirley Chater, Commissioner of SSA
Becky Ogle, Campaign Committee





Patricia Steinitz
314 Richey Avenue
Pittsburgh, PA 15214
(412)-231-1201
(412)-231-3159 Fax

July 24, 1996

Dear Mr. President,

I am writing to you regarding a matter of the utmost concern. You must not pass the Welfare Reform Bill due to the inclusion of the changes it contains regarding children who receive benefits under the Social Security Supplemental Income Program. The elimination of the Individual Functional Assessment will cut off thousands of medically needy children including my daughter, Hunter. Although Hunter does not receive a cash grant, she has a medical card as a result of her disability status. Without her medical card my husband and I would not be able to work and Hunter's health would be drastically effected.

Hunter was born with an extremely rare, life threatening skin disorder called Harlequin Syndrome. It is one of the most rare forms of a family of skin disorders called Ichthyosis. She is one of only 3 surviving children in the U.S. with this disorder. Harlequin Syndrome is passed on genetically and is a lifelong condition.

In brief, Hunter's rate of skin production is extremely accelerated. She has an extremely high caloric need as result of her skin production. All of her food is supplemented with calories and every bite of her food is important. Even with the high caloric diet she still looks like an undernourished child. Hunter's body can not regulate her body temperature due to thickening of her skin which is a result of her skin production. Overheating and inability to maintain fluids subject her to dehydration much more quickly than the average child. Overheating and poor nutrition can contribute to her chance of infection. Her skin disorder alone puts her at risk to skin infections and at risk to sepsis which is the most common cause of death of these children.

At birth Hunter's whole body was covered with very thick scales of skin that obscured her fingers, toes, ears, and nose. Her

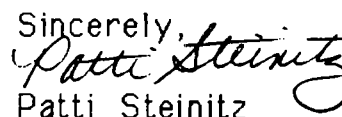
doctors told us that she would probably die. Most children born with this disorder die due to infection and excessive loss of body fluids, protein, and electrolytes. To everyone's amazement, Hunter survived. She is now 21 months old. Her care is complex and time consuming. Aside from her medical care she receives Early Intervention services due to physical problems that are a result of her skin disorder.

Hunter qualified for disability status for SSI under the IFA. I have been told that her disability is not on the "Medical Impairment List" which SSI uses to qualify children. I have been trying to get a copy of the list for over a year and have been unsuccessful. My understanding is that the vast majority of rare disorders are not on the Medical Impairment List. As a result, the only way to qualify for SSI is under the IFA. This is why it is so important to keep the IFA. If it is eliminated thousands of children with rare disorders will be cut off of SSI and will lose necessary benefits.

The most important benefit for Hunter is her medical card. It covers the skilled nursing care that is required to assure continuity of care while my husband and I are at work. The nursing care also allows us to sleep. Without Hunter's medical card my husband and I would be forced to quit our jobs and go on welfare. Without Hunter's medical card her care would decline and leave her at risk to illnesses and death. This is an option I do not wish to pursue.

I understand that Welfare does need reformed. I am a tax payer and do agree with some of the reform. However, I do not believe that thousands of children should suffer in order to reform the rest of the Welfare system. I urge you to consider how the elimination of the IFA will effect so many children and their families. I urge you to veto the Welfare Reform Bill if it contains anything that is detrimental to disabled children.

I am sending via overnight mail some photos of Hunter and an article that was published in our local newspaper. Thank you for your attention to this matter. I look forward to your response to my concerns.

Sincerely,

Patti Steinitz

THE WHITE HOUSE
WASHINGTON

September 3, 1996

Ms. Patricia Steinitz
314 Richey Avenue
Pittsburgh, PA 15214

Dear Ms. Steinitz:

Thank you for your recent letter concerning welfare reform. Your comments are especially helpful as we move forward to implement the new legislation.

Welfare reform presents us with perhaps the most significant opportunity to free Americans from the cycle of poverty. However, signing the legislation was a difficult decision for the President because of the bill's weaknesses. For example, the bill makes deeper cuts than it should in nutritional assistance programs, especially for working families with children. Further, Republican-led efforts to cut off assistance to legal immigrants who work hard, pay taxes, and serve America prevailed. I can assure you though, that this Administration is committed to pushing for changes in these and other provisions so the legislation is responsive to all poor Americans.

Despite some of its shortcomings, I believe this legislation is a significant improvement over the earlier versions the President vetoed. The Congressional leadership was intent on eliminating the guarantee of health care for the poor, the elderly, and the disabled -- and we were able to stop them. In addition, we prevented these same leaders from destroying the food stamp and school lunch programs that provide basic nutrition to working families and poor children, and we worked vigorously to protect child abuse prevention and foster care, and support for young unwed mothers.

The welfare reform legislation gives us an unprecedented chance to provide millions of women and children with a chance to escape welfare as a way of life. For too long, America's poor families have been trapped in a cycle of dependency that fosters a sense of hopelessness and despair.

The real challenge ahead lies in our ability to ensure that states move people from welfare to work by offering on-the-job training, subsidized work and other *real* work options. Transforming the welfare system to one based on work and personal responsibility will empower Americans.

September 3, 1996
Page 2

Again, thank you for providing us with your thoughtful comments and analysis on welfare reform. As the President observed at the signing ceremony, the passage of this legislation marks a beginning, not an end. Now, we must all work together to make sure that families and children are protected as we move citizens from welfare to work.

Sincerely,

A handwritten signature in cursive script, reading "Carol Rasco". The signature is fluid and elegant, with the first name "Carol" and the last name "Rasco" clearly distinguishable.

Carol Rasco
Assistant to the President for
Domestic Policy

The Washington Post

AN INDEPENDENT NEWSPAPER

A Choice for the Administration

THE WELFARE bill that Congress passed and the president signed two months ago had to do with more than just welfare. Among the other programs it will affect is a special form of federal aid to needy families with disabled children. For years a kind of backwater in the budget to which no one paid much attention, the program in the 1990s suddenly more than tripled in size. The caseload shot up from 300,000 to about a million, the cost from a little more than \$1 billion to about \$5 billion.

Partly this expansion reflected a 1990 Supreme Court decision requiring that eligibility standards for children be eased, and partly it was the result of an earlier congressional decision to admit to the program a broader range of children with mental disorders. But critics had begun to contend that it was partly due to abuse as well. They argued mainly on the strength of anecdotal evidence that parents and others were taking advantage of the liberalized rules to obtain disability benefits for children who, while needy, were not in fact disabled, and they said the program had grown in directions that Congress never fully had contemplated.

The House version of the welfare bill therefore proposed sharp cuts in the disability benefits. Rather than give cash to eligible families, it proposed offering most of them only certain services, limiting the cost of those and narrowing the definition of disability. The Senate took a more moderate approach, which prevailed in the bill the president finally signed. The cash payments were preserved, and the disability definition was tightened less.

The new language creates a zone of discretion for the administration. A child will be deemed disabled if he or she has a "medically determinable physical or mental impairment, which results in marked and severe functional limita-

tions." What might those be? The administration will have to say in regulations.

Just about everyone agrees that some three-fourths of the children on the rolls are disabled enough that they will meet the new test, however it is phrased. It is the others, about a quarter of a million, whose fate is up in the air, together with however many might be expected to apply in the future.

Advocacy groups say the final language in the bill was deliberately such that the administration need cut off only a minority of these. They cite, among much else, a carefully orchestrated colloquy on the Senate floor last year when this part of the bill was taking final form. Then-Majority Leader Bob Dole, who took part along with Sens. John Chafee and Kent Conrad, said at one point, "I think we can all agree that the children's SSI [as the program is called] needs a tune-up." That hardly argues for wholesale change, they say. They make the further point that the Social Security Administration has already tightened up on the program, such that the so-called allowance rate or percentage of applicants granted benefits has fallen from an artificial high of 70 percent in 1991 to 30 percent today.

Our own sense is that the administration ought to err on the side of giving the extra benefit rather than denying it. The poverty rate among children remains high, and other forms of aid to needy families with children already are being cut. Having at home a child with even a relatively modest disability can be an enormous burden and cost for even a family of some means, if only because it often deprives the household of earnings. A family member who otherwise might work is required to provide child care instead. We have no doubt that people here and there are ripping off this program, just like any other program. But most aren't, and that ought not be the presumption of policy.

Paul Block, publisher, 1927-1941

Paul Block Jr., co-publisher, 1942-1987

William Block, co-publisher, 1942-1989; chairman, 1990 -

John Robinson Block

Co-publisher and editor-in-chief

William Block Jr.

Co-publisher and president

John G. Craig, Jr., editor and vice-president

Madelyn Ross, managing editor

Michael McGough, editorial page editor

Robert B. Higdon, vice-president and general manager

Setting an SSI standard

Clinton can help families with disabled children cope

Hundreds of thousands of families are in limbo, not sure if the SSI money they receive to help care for their disabled children will continue.

Their fate became tied up with the welfare reform bill that was signed into law over the summer. That bill included a provision to tighten eligibility for Supplemental Security Income, but it left the administration considerable discretion in pinning down the details.

Now the needy families who count on the maximum monthly SSI benefit of \$458 have to wait until the Social Security Administration defines exactly what the bill means when it says a child is disabled if he or she has a "medically determinable physical or mental impairment, which results in marked and severe functional limitations."

The administration could try to work backward by figuring out how many of the nearly 1 million children eligible for SSI should be lopped from the rolls and craft a definition that accomplishes that.

A more humane and reasonable approach would be to come up with a standard that protects the families of children with a legitimate need: to "tune up" the system as the senators who wrote the bill intended, rather than gut it.

SSI for families of disabled children never received much attention until 1990 when a Supreme Court ruling expanded eligibility, which helped triple the caseload at an annual cost of \$5 billion.

Following that explosive growth came charges of broad-based abuse of the system, including accounts of parents coaching children to appear retarded or hyperactive. Numerous investigations into the claims have shown them, at best, to be wildly exaggerated. No broad-based or systemic abuse was uncovered.

Administration has been much more restrained in awarding the benefits. Today, 70 percent of applicants are rejected compared with a 70 percent acceptance rate five years ago.

The House wanted to clamp down much more tightly on the program, but the more moderate Senate approach prevailed. Most of the children receiving SSI will be eligible no matter how the definition is drawn. But that leaves about 250,000 families who don't know whether they will still qualify.

It was that now-defunct assessment that led to SSI eligibility (and therefore Medicaid eligibility) for 23-month-old Hunter Steinetz of Observatory Hill, whose disability is so rare that it is not listed on the standard directory of qualifying medical conditions. Hunter's skin grows so fast that her body cannot shed it quickly enough. Without costly and exhausting around-the-clock medical treatment, Hunter will not survive.

Other children ruled eligible under the old assessment may have had several problems, each of which on their own would not be enough to constitute a severe disability. The old standard allowed consideration of the cumulative effect. Those families may not need the hundreds of thousands of dollars of care that Hunter requires. But they may spend thousands of dollars in special transportation or clothing or nutrition or day-care costs that they can't afford on their own.

The Social Security Administration should not merely rubber-stamp what was done in the past. But in defining what constitutes "marked and severe functional limitations," it should strive to remember that this is one of those areas in which government can and must be part of the solution. Its role here is not to save money at any cost but to help deserving families with disabled children cope.

In the meantime, the Social Security

OCTOBER 10, 1996

BILL CLINTON'S GENDER C



LETTER

Churches face many ob.

Recently, Mayor Murphy spoke to church leaders and scolded them ("Churches Urged to Fight Crime," Sept. 28) for not doing enough to address the problems of the urban population. I can appreciate his frustrations, but he is directing his anger at the wrong people. Church leaders in Pittsburgh are faced with the same problems as city leaders. We have an older population. The numbers attending church have dwindled and finances are tight.

Murphy's expectations of church leaders come at a time when we no longer have the power or influence he would like us to have. Supreme Court rulings have taken away religious influence from public schools, graduation ceremonies and Christmas displays on public property. Too, there's fierce opposition to giving tax money to parents who want to send their children to denominational schools.

Other obstacles to the church's role in society include the fact that religious-based charities may not receive government funds. Another is that worshipers are denied access to their churches when there is a marathon in the city. And of course,

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TOM DASCHLE
SOUTH DAKOTA

United States Senate
Office of the Democratic Leader
Washington, DC 20510-7020

October 4, 1996

The President
The White House
Washington, D.C. 20500

Dear Mr. President:

You have an opportunity to implement the recently enacted welfare reform legislation in a manner that treats low-income disabled children fairly. In crafting a new definition of disability for children under the Supplemental Security Income (SSI) program, Congress provided the executive branch with great latitude to interpret the statute. Knowing of your long-standing commitment to these children, I know you will use that latitude wisely.

My staff and I were deeply involved in crafting with Senator Dole, Senator Chafee and Senator Conrad the compromise language that ultimately became the basis for the new law. We made a conscious and sustained effort to ensure that the Social Security Administration was granted considerable discretion to implement regulations that would tighten the program without dropping truly disabled children from the rolls. This understanding is confirmed by the views of the Congressional Budget Office (CBO) at the time; CBO told Congress that the new policies could cut between 10 to 28 percent of the children from the program, depending upon SSA's regulatory interpretation.

A great deal of effort went into forging a bipartisan compromise over reforming this program. In the end, we reaffirmed that a functional assessment of a child's abilities was critical in evaluating childhood disability. The legislative history makes clear that, to accomplish this, SSA should establish a functional assessment beyond the "Listings of Impairments." The new definition of disability, requiring that qualifying impairments be "marked and severe functional limitations," explicitly does not establish the listings level of severity, or any equivalent measure, as the basis for determining childhood disability. For SSA to interpret the statute otherwise would be a tragic mistake with potentially devastating consequences for thousands of this nation's most vulnerable children.

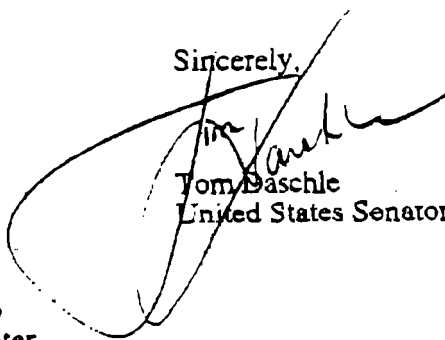
Certainly, the new statute requires SSA to eliminate the old Individualized Functional Assessment. It does not, however, compel SSA to adopt the very strict level of the listings. A better approach, which we envisioned when crafting the compromise language, would require one marked and one moderate disability in order to qualify. This approach is supported by several respected organizations representing children with disabilities with whom we consulted in the process of developing the new definition. Such an approach meets the statutory requirement that the test determine eligibility only for "marked and severe functional limitations" without requiring the listings level of severity.

October 4, 1996
Page Two

I trust that you will do everything you can to strike a balance that ensures only those children who are severely disabled receive SSI benefits, without denying those who are truly deserving. Thank you for your consideration of this legislative history in interpreting the new law in the best interest of America's most vulnerable children.

With best wishes, I am

Sincerely,

A large, stylized handwritten signature in black ink, which appears to read "Tom Daschle". The signature is written over the printed name and title.

Tom Daschle
United States Senator

cc: The Honorable Carol Rasco
The Honorable Shirley Chater

CHAIRMAN, COMMITTEE ON
ENVIRONMENT AND PUBLIC WORKS
COMMITTEE ON FINANCE
JOINT COMMITTEE
ON TAXATION
SENATE ASKS CONTROL
OBSERVER GROUP

United States Senate

WASHINGTON, DC 20510-3902

September 17, 1996

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SENATE@SENATE.Senate.gov

The Honorable Bill Clinton
President of the United States
The White House
Washington, DC 20500

Dear Mr. President:

Your administration has a key role to play in the implementation of the children's Supplemental Security Income (SSI) provisions that were included in the welfare reform bill enacted last month. While we are all interested in ensuring that only children who are truly disabled receive SSI benefits, we are equally concerned that those children who are, in fact, severely disabled remain eligible for the program. The Social Security Administration (SSA) has the difficult responsibility of striking a balance between these two goals.

The statutory language was intended to give SSA substantial discretion in drawing the eligibility line for this program. Clearly, the new law cannot be read to allow SSA to continue the current level of severity which drew so much criticism. At the same time, the new definition was never intended to "gut" the program and, in fact, affirms the importance of functional assessment as part of an effective evaluation of childhood disability.

The debate over this issue was heated at times, but, ultimately, we reached a compromise on the definition of childhood disability in September, 1995. That definition became part of the overall Congressional compromise on SSI, and was included in the first two versions of welfare reform approved by Congress and then finally in the bill enacted in August. The compromise is notable in two ways. First, it preserves a broad functional approach beyond the "Listings of Impairments," in measuring childhood disability. Second, it specifically does not establish the listings level of severity, or any equivalent level of severity, as the measure to be used in assessing childhood disability.

The enclosed Senate colloquy between those of us involved in this compromise is important in understanding the meaning of the new definition. This colloquy was not entered into lightly. Rather, it was the subject of much negotiation and was key to the final language of the definition regarding "physical and mental impairment, which results in marked and severe functional limitations" after dropping the requirement that the effect of the impairment also be "pervasive".

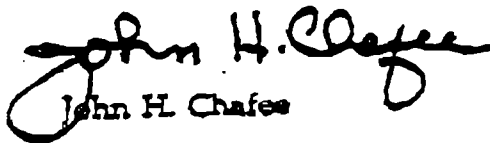
The Honorable Bill Clinton
September 17, 1996
Page two

> It is certainly appropriate for SSA, as the regulatory agency, to adopt a disability test that is stricter than the old Individualized Functional Assessment (IFA), but which is not at the very strict level of the "Listings." The proposal put forward by several disability advocates and organizations with considerable expertise - a one marked/one moderate level - is an acceptable and reasonable approach that fulfills the statutory demand for a test that allows benefits only for marked and severe functional limitations, but does not require that these limitations be pervasive.

The Congressional Budget Office (CBO) has also acknowledged that SSA would have a great deal of flexibility in meeting the requirements of the new law. The enclosed Senate Finance Committee report shows that CBO estimated that the new definition of childhood disability could bar anywhere from 10-28 percent of children from the program, depending upon the regulatory interpretation of the new definition.

I know that you will do everything in your power to ensure that children with severe disabilities who are truly deserving are not harmed by the changes in the new welfare law. Thank you in advance for your attention to this matter. Please do not hesitate to contact me if I may be of any further assistance.

Sincerely,


John H. Chafee

JHC:bd

cc: Secretary Shalala
Commissioner Chater

There are several subtypes of the disease. Acute eruptive lichen planus is probably the most common form in children. The lesions erupt in an explosive fashion, much like a viral exanthem, and spread to involve most of the body surface. Hypertrophic, linear, bullous, atrophic, annular, follicular, erosive, and ulcerative forms of lichen planus may also occur. Nail involvement may occur in the chronic forms but is rarely evident in children (twenty nail dystrophy, Sec. 24.22). The disorder may persist for months to years; the acute eruptive form is most likely to involute permanently. Frequently, intense hyperpigmentation persists for a long time following resolution of lesions. The pathology of lichen planus is quite specific, and a biopsy is indicated if the diagnosis is unclear.

Treatment is directed at alleviation of the intense pruritus as well as amelioration of the skin lesions. Oral antihistamines and/or tranquilizers are often helpful. The skin lesions respond best to regular applications of a topical corticosteroid preparation. Rarely, systemic corticosteroid therapy is necessary to gain control of widespread, intractable lesions.

Porokeratosis is a rare, chronic, progressive disease that is inherited as an autosomal dominant trait. Several forms have been delineated: solitary plaques, linear porokeratosis, hyperkeratotic lesions of the palms and soles, disseminated eruptive lesions, and superficial actinic porokeratosis. The last form, probably induced by excessive sun exposure, occurs more commonly in adult females. Other types of porokeratosis are more common in males and begin during childhood. Sites of predilection are the limbs, face, neck, and genitalia. The primary lesion is a small, keratotic papule that enlarges peripherally so that the center becomes depressed, the edge forming an elevated wall or collar. The configuration of the plaque may be round, oval, or gyrate; its elevated border is split by a thin groove from which minute cornified projections protrude. The enclosed central area is yellow, gray, or tan, sclerotic, smooth, and dry, whereas the hyperkeratotic border is a darker gray, brown, or black.

The differential diagnosis includes warts, epidermal nevi, lichen planus, granuloma annulare, and elastosis perforans serpiginosa. A skin biopsy will disclose the characteristic cornoid lamella (plug of stratum corneum cells with retained nuclei) which is responsible for the linear ridge of the lesion, an invariable clinical feature.

The disease is slowly progressive but relatively asymptomatic. Lesions are sometimes responsive to applications of liquid nitrogen or may be surgically excised, a procedure which may not be feasible.

Papular acrodermatitis of childhood (Gianotti-Crosti syndrome) is a distinctive eruption associated with malaise and low-grade fever but few other constitutional symptoms. The incidence peaks in early childhood. Occurrences are usually sporadic, but epidemics have been recorded.

The skin lesion is a monomorphous, usually nonpruritic, dusky or coppery red, flat-topped, firm papule ranging in size from 1 to 5 mm. The papules appear in crops and may become profuse but remain discrete forming a symmetrical eruption on the face, buttocks, and limbs including the palms and soles. At times the papules become hemorrhagic. Lines of papules (Koebner phenomenon) may be noted on the extremities. The trunk is relatively spared, as are the scalp and mucous membranes. Generalized lymphadenopathy and hepatomegaly constitute the only other abnormal physical findings. The eruption resolves spontaneously in about 3 wk. Lymphadenopathy and hepatomegaly may persist for several months.

This eruption is most often associated with primary infection with hepatitis B virus and is, therefore, associated with hepatitis B surface antigenemia. Subtyping of the HB_{Ag} in most instances has demonstrated the determinant *ayw*. Elevation of serum transaminase and alkaline phosphatase values

without concomitant hyperbilirubinemia is usual, and histologic changes of viral hepatitis are demonstrable on liver biopsy. Skin biopsy is characterized by a perivascular mononuclear cell infiltrate and capillary endothelial swelling.

Generally the disease is benign and self-limited and does not recur. The hepatitis usually resolves in 2-3 mo but, occasionally, may progress to chronic hepatitis with persistent antigenemia and elevated transaminase activity. The surface antibody of the hepatitis B virus is not detected during the phase of dermatitis appearing approximately 6-12 mo later in patients who become HB_{Ag} negative.

A similar cutaneous eruption has also been observed in patients infected with Epstein-Barr virus, coxsackievirus A16, and parainfluenza virus and undoubtedly accompanies other viral infections. Papular acrodermatitis can be confused with lichen planus, erythema multiforme, histiocytosis X, and Henoch-Schönlein purpura.

24.16 ICHTHYOSIS

The ichthyosiform dermatoses are a group of inherited keratinizing disorders characterized by visible scaling in distinctive patterns of distribution. They are usually distinguishable on the basis of inheritance patterns, clinical features, associated defects, and histologic changes. Since some of these conditions cause disfigurement and considerable mental anguish, early diagnosis is helpful in order to predict probable course and prognosis and to provide supportive management for the patient and family.

Lamellar ichthyosis is a very rare keratinizing disorder that is inherited as an autosomal recessive trait. Affected infants are extremely grotesque. Markedly thickened, ridged, and cracked skin forms horny plates over the entire body, disfiguring the facial features and constricting the digits. Severe ectropion and chemosis obscure the orbits; the nose and ears are flattened, and the lips are everted and gaping. Nails and hair may be absent. Joint mobility is restricted, and the hands and feet appear fixed and ischemic. The infants have respiratory difficulty and suck poorly. Most succumb within the 1st wk of life; few live beyond 6 wk. The prognosis is hopeless. Prenatal diagnosis has been accomplished by fetoscopy and fetal skin biopsy.

The **collodion baby** is covered at birth by a thick, taut membrane resembling oiled parchment or collodion, which is subsequently shed. The condition is usually a primary manifestation of one of the ichthyoses, most often of the lamellar variety. Infrequently an affected infant has normal skin after the membrane is shed. There is ectropion, flattening of the ears and nose, and fixation of the lips in an O-shaped configuration (Fig. 24-31). Hair may be absent or may perforate the horny covering. The membrane cracks with initial respiratory efforts and, shortly after birth, begins to desquamate in large sheets. Complete shedding may take several weeks, and occasionally a new membrane may form in localized areas.

The nursery course may be complicated by cutaneous infection with yeasts and bacteria. The outcome is uncertain, and accurate prognosis is impossible with respect to the subsequent development of ichthyosis. Maintenance in a high-humidity environment and application of nonocclusive lubricants may facilitate shedding of the membrane.

Ichthyosis vulgaris, the most common type of ichthyosis, is transmitted as an autosomal dominant trait. Onset occurs sometime after the first year of life. Scaling is most prominent on the extensor aspects of the extremities and back. The flexures are spared, and the abdomen and face are relatively uninvolved. Accentuated markings and creases are apparent on palms and soles. Atopy is relatively common.

The histologic changes differ from those of other types of

Nursing a Special Patient: The Infant with Congenital Ichthyosis—Harlequin Syndrome

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WHEN OUR NICU was suddenly presented with a "harlequin fetus," his initial appearance and requisite care seemed almost overwhelming. A search of the nursing literature yielded no references, and an extensive review of the medical literature yielded only ten case studies of this extremely rare syndrome.¹⁻⁵ We were faced with an unusual and difficult nursing challenge with scanty resources to help us plan care. This article outlines some of the strategies we used to provide care for this very special infant and his family.

Harlequin fetus is considered the most severe form of the congenital ichthyosiform dermatoses.¹ Its primary feature is thickening of the stratum corneum of the skin. Analysis of the skin from different infants with this condition has revealed abnormalities in protein content, cholesterol, and triglycerides, suggesting that the disorder may have several causes.^{2,3}

The skin appears waxy and yellow and is formed into thick hexagonal plates that are divided by deep erythematous fis-

tures, giving the appearance of reptilianlike scales. The scale-like plates or plaques tend to obscure facial features, so the ears may not be visible, the nose reduced to only the nasal openings, the eyes obscured by everted, contracted lids (ectropion) and edematous conjunctiva, and the mouth contracted and fixed in a piscine appearance (ectlabion). Body mobility is severely restricted, and the infant may be fixed in a single posture (Figure 1).

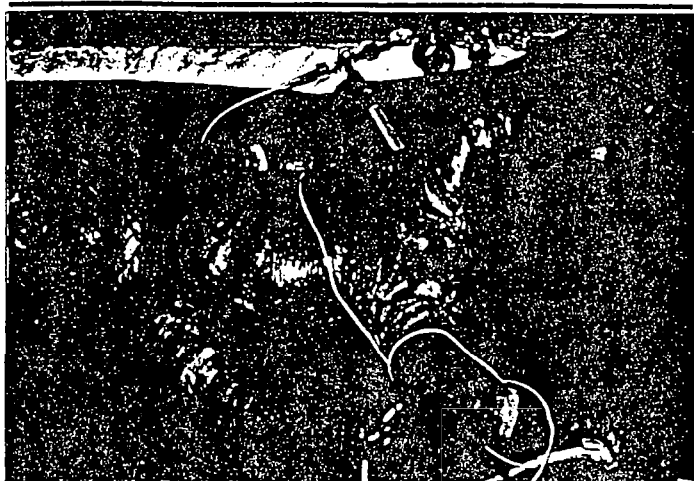
Congenital ichthyosis is thought to be a rare, autosomal recessive trait, which in its most severe form is incompatible with life.⁴ Death usually occurs as a result of overwhelming sepsis due to the loss of skin integrity between the thickened plaques.

OUR INTRODUCTION TO THE INFANT

As is typical, our infant was not diagnosed prenatally. Delivered by cesarean section because of breech presentation,

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FIGURE 1 ■ Day 1: Anterior view of entire body. Note extensive fissuring of abnormal skin, rigid body posture, and thick covering over hands and feet.



Joey (not his real name) arrived in the NICU accompanied by the infant resuscitation team. His scalelike skin was covered with meconium, his eyes occluded by edematous conjunctiva, and his mouth and nose contracted and everted (Figures 2 and 3). His appearance bore no resemblance to a 37-week, 2,530 gm human infant. It was distressing to the professional staff, but it was far more distressing to the first-time parents, who had eagerly anticipated his birth.

Initially, Joey's physiological problems were respiratory because the large plaques of skin restricted chest expansion. Every respiratory movement caused the plates to split, revealing subcutaneous tissue in each fissure. Monitoring him posed problems because electrodes could not be placed effectively on the abnormal skin. Soon after stabilization, we focused attention on protective isolation, hydration, nutri-

FIGURE 3 ■ Day 1: Lateral view of face and head. Note occlusion of ear canal with abnormal keratin and apparent absence of pinna. Hair appears absent but grows within the fissures between plaques.

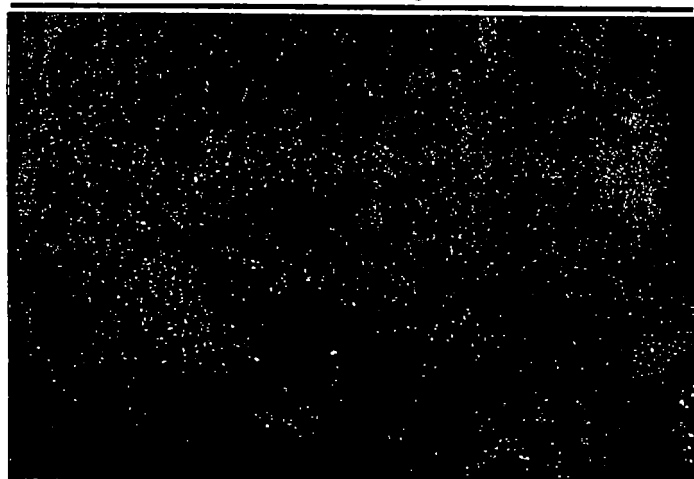


tion, and skin care. The baby was clearly uncomfortable.

Nursing care of the parents was directed at supporting them in their grief response to the loss of the expected normal child and preparing them realistically for Joey's future. Inherent in this support was the effort to create as much normalcy as possible within the existing situation.

The medical treatment for this baby was limited to antibiotics, an ocular lubricant to protect the corneae, oral etretinate (a derivative of vitamin A that had been used successfully in at least one documented case of ichthyosis to promote a more normal pattern of cell growth in the skin), and an ointment to reduce fluid loss from the defective skin.⁵ Several antimicrobial ointments were tried, but sterile Vaseline worked best. This was the treatment of choice.

FIGURE 2 ■ Day 1: Anterior view of face and head. Note ectropion and ectlabion (marked) and obliteration of facial features: conjunctiva severely edematous, occluding normal eyes; eyelids unable to close; mouth rigid and unable to close.



ORGANIZING JOEY'S CARE

Basically, survival of the baby and of the family unit depended on expert nursing care—especially in creating a safe, comfortable environment for Joey. Because the infant's appearance was so bizarre (many nurses felt it "hard to believe that he really was a baby"), a core group of nurses with an interest in caring for him was formed. This core group, representing nurses from all shifts, provided consistency and continuity of care for Joey. The limited number of nurses meant that Joey's parents had familiar caregivers to interact with and that Joey's nurses developed expertise in his skin care regimen. A complex and time-consuming routine, skin care greatly interfered with Joey's rest. Having experienced nurses perform it significantly reduced the time factor.

Nursing staff initiated a contact with Kingston General Hospital (Kingston, Ontario) after telephone queries to all major perinatal centers in Ontario found it to be the only NICU with experience in nursing this type of patient. A multidisciplinary team conference was organized to discuss

how the team could best meet the needs of Joey and his stricken parents.

Based on these discussions and on the available medical literature, we developed a comprehensive nursing care plan that addressed six major nursing diagnoses (see Table 1):⁶

1. Potential for infection
2. Ineffective thermoregulation
3. Impaired physical mobility
4. Potential fluid/nutritional deficit
5. Alteration in comfort
6. Alteration in parenting

SPECIAL PROBLEMS

One of the unique problems that an infant with harlequin syndrome poses to nurses is that of maintaining confidentiality. Although one can appreciate the desire of professional students and staff to see a patient with a rare anomaly, restricted access is in the patient's best interest. Joey's visitors were limited to those professionals directly involved in his care, and his incubator was screened to protect him from the curious. The screens were removed once his skin began to look more normal.

Joey presented some additional challenges: Infant servo-control probes could not be taped to his skin, axillary thermometers could not be used to monitor his body temperature, limb band electrodes were required for cardiac monitoring, and blood gases were the only reliable means of monitoring his oxygen and carbon dioxide levels. Umbilical arterial and venous lines were essential because peripheral vessels were not visible until his skin improved. He required tube feeding because his mouth was too rigid to allow him to suck, and impaired skin integrity and stress raised his caloric needs. All his special needs meant that he required one-to-one nursing care for the first month of his life.

Skin Care

Skin care, initially performed using sterile technique, closely resembled that given to patients with extensive burns. Joey required the high, heated humidity available only in an incubator because he was unable to maintain his body temperature, and moisture prevented some of his skin from cracking. The humidity helped to reduce evaporative heat loss from the weeping fissures.

The humidification system used was external and sterile, used sterile water, and was exchanged every 24 hours. Because every effort was made to maintain a closed system, we do not believe that the subsequent development of sepsis originated from this source. The causative organisms of the sepsis were consistent with those of integumentary and enteric origin and not those normally associated with humidification systems.

Joey's hands and feet were encased in thick skin, like mittens, that impaired circulation to his digits. Because he had some necrosis of tissue in his fingers, at one point, as concern over the condition of these extremities rose, fasciotomy was

FIGURE 4 ■ Day 23: Anterior view of entire body. Note marked improvement in skin (dark areas are remains of old plaque); Thick "mittens" still present on hands and feet. Thick plaques still present on top of head. Note flexion of limbs.



considered and abandoned. Instead, we instituted conservative treatment using complex moist dressings. The "mittens" split and sloughed naturally with no permanent damage to the fingers or toes. Skin care for Joey took approximately 8 to 12 hours over a 24-hour period.

Gradually, the platelike scales split and peeled off, revealing shiny, erythematous skin. Scales reformed but gradually diminished in thickness, and the skin color faded from yellow to pale and translucent (Figure 4).

Throughout his short life, Joey exuded a strange odor that one nurse described as "fungal" in nature. Despite scrupulous skin care efforts, which were impeded by the nature of the skin and by the distress cleansing caused the infant, the odor persisted. It was probably caused by a mixture of decomposing meconium, necrotic tissue, and *Candida* colonization. This odor was a distressing part of nursing Joey.

While Joey's skin was changing, his eyelids gradually improved, and he began to respond to visual cues. He appeared to have normal vision. Similarly, mobility in his joints increased, though he never achieved a normal range of motion. He had limited movement of his head and neck. Necrotic areas on the tips of his fingers and toes healed as the plaques sloughed and Joey began to move. In short, with care, a human infant was emerging from his reptilian cocoon (Figures 5 and 6).

PARENTAL SUPPORT

Nursing staff supported Joey's parents in their initial introduction to him and encouraged them to express their feelings verbally. Their method of coping was to engage in information seeking, which the nurses aided by providing honest answers to all questions and by directing them to other sources of information. They gradually became lay experts on congenital ichthyosis.

From the beginning, the parents were encouraged to participate as much as possible in Joey's care and to hold and cuddle him as his condition stabilized. Even though his prognosis was poor, the nursing goal was to establish as much nor-

TABLE 1 ■ Nursing Care Plan for Infant with Congenital Ichthyosis

Nursing Diagnosis	Goal	Intervention
I. Potential for infection related to: Impaired skin integrity Developmental stage (neonate) Invasive procedures such as umbilical line Increased environmental exposure	Infection will be identified early. Sepsis will be prevented.	1. Use aseptic technique. Assure sterile sheets, diapers, feeding tubes, formula, baths. Change incubator q 24 hours. Use reverse isolation: gowns, gloves, hat, mask. 2. Take vital signs q 2 to 4 hours. 3. Assess open areas q shift. 4. Send samples from skin and eyes q 3 days for culture and sensitivity. 5. Cleanse eyes with 0.9% NaCl q 3 hours; apply ointment (Lacrilube). Change respiratory equipment q 24 hours. Minimize access to IV lines; cleanse ports with betadine and alcohol. 6. Cleanse skin with sterile, warm NaCl 0.9%. 7. Apply copious amounts of sterile Vaseline to sloughing plaques. 8. Minimize handling. 9. Clean perineum well with NaCl 0.9%. Apply nystatin ointment to fissures in perineum. 10. Dress fingers with sterile gauze soaked in sterile Vaseline. 11. As skin fissures heal and plaques slough, change to clean technique.
II. Ineffective thermoregulation related to: Wet body surface Neonate Impaired skin integrity Fluctuating environmental temperature	Temperature will remain between 36.5 and 37° C.	1. Use servocontrol when unstable. 2. Assess temperature rectally prior to handling. 3. Warm linen and diaper. 4. Warm sterile saline prior to bath. 5. Warm salves before application. 6. Warm feedings 20 minutes in incubator before feeding. 7. Use no direct heat application. 8. Cover portholes. 9. Keep from drafts. 10. Provide care and treatments in incubator if possible. 11. Use radiant warmer for outside treatments and parent visits. 12. Provide high humidity to reduce evaporation. 13. Change sheets as soon as they become wet from weeping fissures. 14. Change diapers q 3 hours. 15. Apply Vaseline to all body surfaces.
III. Impaired physical mobility related to: Pain Rigid, thick skin Protective restraints Contractures	Infant will achieve painless, independent, functional movement in all limbs.	1. Give analgesic as required. 2. Cluster treatments around peak times for analgesic. 3. Provide functional positioning with blanket rolls to support joints. 4. Change position q 3 hours. 5. Limit restraints to limbs that can reach umbilical line. 6. Provide range-of-motion exercise to moving joints; increase as plaques slough.

TABLE 1 ■ Nursing Care Plan for Infant with Congenital Ichthyosis (continued)

Nursing Diagnosis	Goal	Intervention
IV. Alteration in comfort related to: Multiple open areas Exposed nerve endings Fatigue Painful treatments Rigid body posture Inability to self-soothe Complex care	Infant will achieve a minimum of 10 hours sleep q 24 hours. Infant will show a relaxed facial expression and body posture. Infant will tolerate wake state, skin care, and handling without crying.	1. Give morphine IV drip 2.5 mg in 100 ml D ₅ W, 1 ml/hour (started on day 3). 2. Give morphine up to 6 ml/hour plus 7 ml/hour bolus prior to skin care. 3. Provide pacifier pm. 4. Position for comfort in infant chair with limb support. 5. Decrease stimuli in environment: Screen incubator, dim lights, place in quiet corner. 6. Decrease tactile stimulation. 7. Schedule care to provide frequent rest periods. 8. Control traffic near incubator.
V. Potential fluid/nutritional deficit related to: Rigid mouth tissue Impaired sucking and swallowing Gagging when fed Increased fluid loss from skin	Fluid balance and weight gain will be achieved. Oral feeding and adequate caloric intake will be established. Blood glucose levels will be within normal limits.	1. Give in-and-out gastric feeds, passing the tube with each feed, until infant is able to feed by mouth. 2. Feed expressed breast milk. 3. Attempt to bottle feed with premie nipple when lips soften and can be approximated. 4. Support mother in attempt to breastfeed when baby can suck (one attempt at age 3 weeks was unsuccessful). 5. Burp frequently because of excessive air ingestion during feeding. 6. Monitor urine specific gravity. 7. Weigh daily. 8. Monitor oral mucous membranes for hydration. 9. Weigh diapers. 10. Provide humidity in incubator. 11. Monitor IV lines and fluid intake.
VI. Alteration in parenting related to: Interruption in bonding Physically and visibly abnormal baby Potential death of infant Odor of baby	Family unit will remain intact, cope with situation, and engage in parenting behaviors.	1. Support family in acute grief response to loss of expected normal infant. 2. Encourage parents to be part of care and decision making. 3. Include parents in team meetings concerning baby's care. 4. Keep parents informed of changes in baby's status. 5. Encourage touching and holding. 6. Role model response to baby by holding and cuddling him in presence of family. 7. Call baby by name. 8. Teach parents to recognize cues that infant is giving, and encourage them to respond to cues. 9. Encourage extended family members to visit. 10. Use photographs to illustrate baby's improvement. 11. Inform parents of other available resources for help, such as social services and psychiatry (not utilized). 12. Give parents opportunity to verbalize fears, concerns. 13. Use core nurses to promote therapeutic relationships.

FIGURE 5 ■ Day 23: Anterior view of head and face. Note improvement in ectlabium (able to suck) and extropion. Thickened skin still present on fingers, digits still not mobile. Thick plaque remains on scalp area.



malcy as possible for both parents and baby while maintaining a realistic view of the infant's future. Nursing staff held, cuddled, and talked about Joey in ways that demonstrated their acceptance of him as an infant. By two weeks, his parents were able to hold him; by three weeks, his mother made an unsuccessful attempt to breastfeed him; by four weeks, extended family members were invited to the NICU.

At the age of five and one-half weeks, Joey contracted a massive gram-negative septicemia. Blood cultures grew *Enterococcus fecalis*, *Enterobacter cloacae*, Enteric streptococcus group 17, Group B streptococcus, and *Streptococcus viridans*. We began aggressive treatment, and medical staff met with the parents frequently to discuss this grave turn of events.

After much deliberation, the parents, physicians, and nurses decided jointly to limit intervention to palliative measures. All

blood work, antibiotics, transfusions, and etretinate were discontinued, and a "Do Not Resuscitate" order was instituted. Joey succumbed to sepsis at the age of six weeks and four days. His parents had successfully assumed a parenting role under very difficult circumstances and had come to know, love, and relinquish their baby son.

EVALUATION OF THE CARE

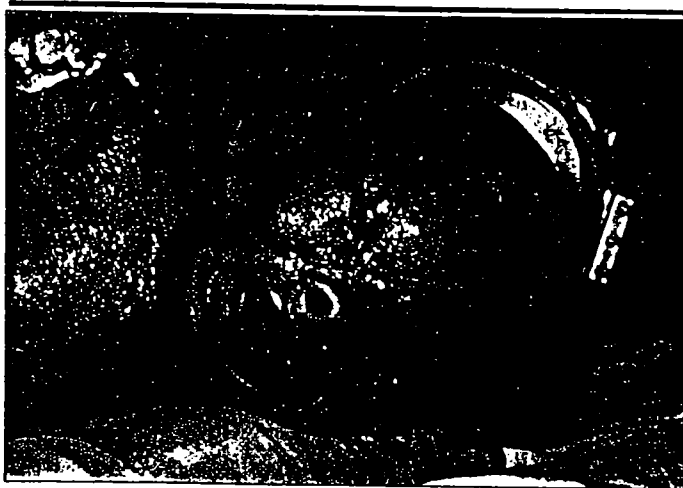
Retrospectively, the concept of aggressively treating an infant with such a poor prognosis may seem futile, but there were substantial benefits to this course of action. From a nursing standpoint, Joey's emerging normalcy eased the task of caring for him because there were visible results of attentive nursing care. We could see the difference we were making in his appearance. It also made it easier for members of the nursing team to support each other. We were engaged in a common goal of assisting in the development of both Joey and his family.

Joey's short life with his improved physical appearance gave his parents time to accept his condition and accept him as their son. The time also provided them the opportunity to contact existing resources and explore what is known about harlequin syndrome and its sad prognosis. This information allowed them to make decisions about their baby's care in an atmosphere of informed consent and to develop a solid therapeutic relationship with the treatment team.

All these factors facilitated the process of normal grieving, allowing the couple time to acquire and develop supports that helped them in their loss. The decision to withdraw aggressive treatment and give palliative care was a difficult one for all participants—professionals and parents alike. Some of the nurses, for example, had begun to hope for more of a future for Joey than realistically was possible.

It is probably more difficult to establish meaning for this experience for Joey, and this factor may have influenced the

FIGURE 6 ■ Day 23: Anterior head (right side). Note normal ear structure and appearance of remnant of abnormal skin on hand.



CALL FOR ABSTRACTS

Mead Johnson Nutritionals, in continuing their support of perinatal nursing, is pleased to announce the creation of a forum to showcase research in perinatal nursing.

The first annual invitational forum will be held at the Marriott's Mountain Shadows Resort, Scottsdale, Arizona, January 24-27, 1992. Faculty for the forum are Maureen Keefe, PhD, RN, and Paula Meier, DNSc, RN.

This is a call for abstracts. For abstract form and application call (812) 429-5839 or see your Mead Johnson Nutritionals representative. Application deadline is September 1, 1991. The 20 participants selected for this meeting will have all expenses paid.

final decision not to continue with aggressive treatment. Beginning on the third day of his life, analgesics had been prescribed because Joey demonstrated pain by crying on movement, by irritability, and by decreasing the amount of time he spent in a sleep state. Physically, the child had multiple areas of raw tissue with exposed nerve endings, so he was never really able to enjoy physical contact. His most comfortable position was sitting semi-erect in an infant seat with blanket rolls to support his stiff joints. In this posture, he was able to sleep for the longest periods. Analgesia ranged from oral and intravenous morphine to chloral hydrate and acetaminophen. Narcotic analgesia was required most of the time to keep him comfortable.

At the time of his death, most of the thick plaques of skin had been shed, Joey was able to suck on a pacifier, and his eyes were almost normal in appearance. In a photograph with his parents, Joey looked almost like any healthy infant. Had we not put so much effort into his skin care, this picture would not have been so meaningful to them now or in the future.

CONCLUSIONS

Our nursing approach reflects how much caring was given to Joey and to his parents by the treatment team and how excellent nursing care transformed a crisis into a more manageable human problem. Although we were unable to affect a long-term positive outcome for the baby, we believe that our care provided a therapeutic environment for this fragile family unit and a basis of support for problem resolution. We were able to successfully address all our nursing diagnoses, with the exception of preventing overwhelming sepsis.

Joey's improved physical appearance represented only one achievement of our nursing care. Of equal importance in treatment success was his parents' acceptance of Joey as their child and their ability to parent to their fullest potential. The decision not to resuscitate Joey was validated by the final autopsy report that listed causes of death: congenital ichthyosis, sepsis, and disseminated intravascular coagulation. Clearly, he could not survive against such overwhelming odds, and their final act of parenting, although extremely difficult, was a judicious one for little Joey. ⑤

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Kevin de Merchant is a graduate of Conestoga College, Kitchener, Ontario, Canada. Since completion of his diploma in 1987, he has worked in neonatal intensive care and pediatric emergency care. Currently employed by the Hospital for Sick Children in Toronto, Kevin was a staff nurse in the NICU at Mount Sinai Hospital at the time of writing and was one of the core nurses caring for the baby.

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The authors wish to thank Maureen Cuddy, Susan Turnbull, and Valerie Wong, who were among the core nurses caring for the infant.

NEONATAL NURSE CLINICIAN/ PRACTITIONER PROGRAM

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Feature Article

INFLUENCE OF NUTRITION ON GROWTH AND DEVELOPMENT OF A LONG-SURVIVING HARLEQUIN FETUS

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ABSTRACT

Two patients with harlequin ichthyosis followed very different clinical courses. One experienced the usually expected early neonatal demise. The other required assisted ventilation for 5 days, but survived thereafter and was alive at the time of report at 5½ years. At 15 months, failure to thrive or develop led to nutritional investigation, which indicated that inhibition of growth was a consequence of protein malnutrition that was a consequence of enormous losses of protein in desquamated skin. Institution of a very high intake of protein led to satisfactory growth and development.

KEY WORDS: harlequin fetus, harlequin ichthyosis, protein nutrition, growth and development

The harlequin fetus is a descriptive term for a neonatal congenital ichthyosis that is generally considered to be distinct from other forms of ichthyosis. First described in 1750,¹ the picture is that of hyperkeratosis and desquamation that form triangular shaped, waxy plaques, reminiscent of the costumes of performers in medieval dramas. Ectropion of each eye and eclabion of the mouth contribute to the striking appearance. In the past, the disorder was considered uniformly fatal in the perinatal period.^{2,3} Recently, however, several survivors have been reported,⁴⁻⁸ two of whom were 2 years of age²⁻⁴ and one 9 years of age.

A patient with harlequin ichthyosis had survived for 5½ years at the time of this report. Unusual protein nutrition was important in his growth and

development. Another patient had the more characteristic neonatally lethal course.

CASE REPORTS

Patient No. 1

A 2.2-kg girl with the characteristics of a harlequin fetus was born to a 24-year-old, gravida 2 para 1 woman. Apgar scores were 8 at 1 minute and 9 at 5 minutes. The infant received oxygen and suction, but was not in respiratory distress. Her skin was pink but scaly and thick. Her temperature was 95.3°F rectally, and she was wrapped in a blanket. She was transferred to the special care nursery of the UCSD Medical Center.

On examination (Fig. 1), the skin had thick scales and was fissured in the areas of flexion. The fishmouth appearance of the mouth was a consequence of contractures around the face. There was

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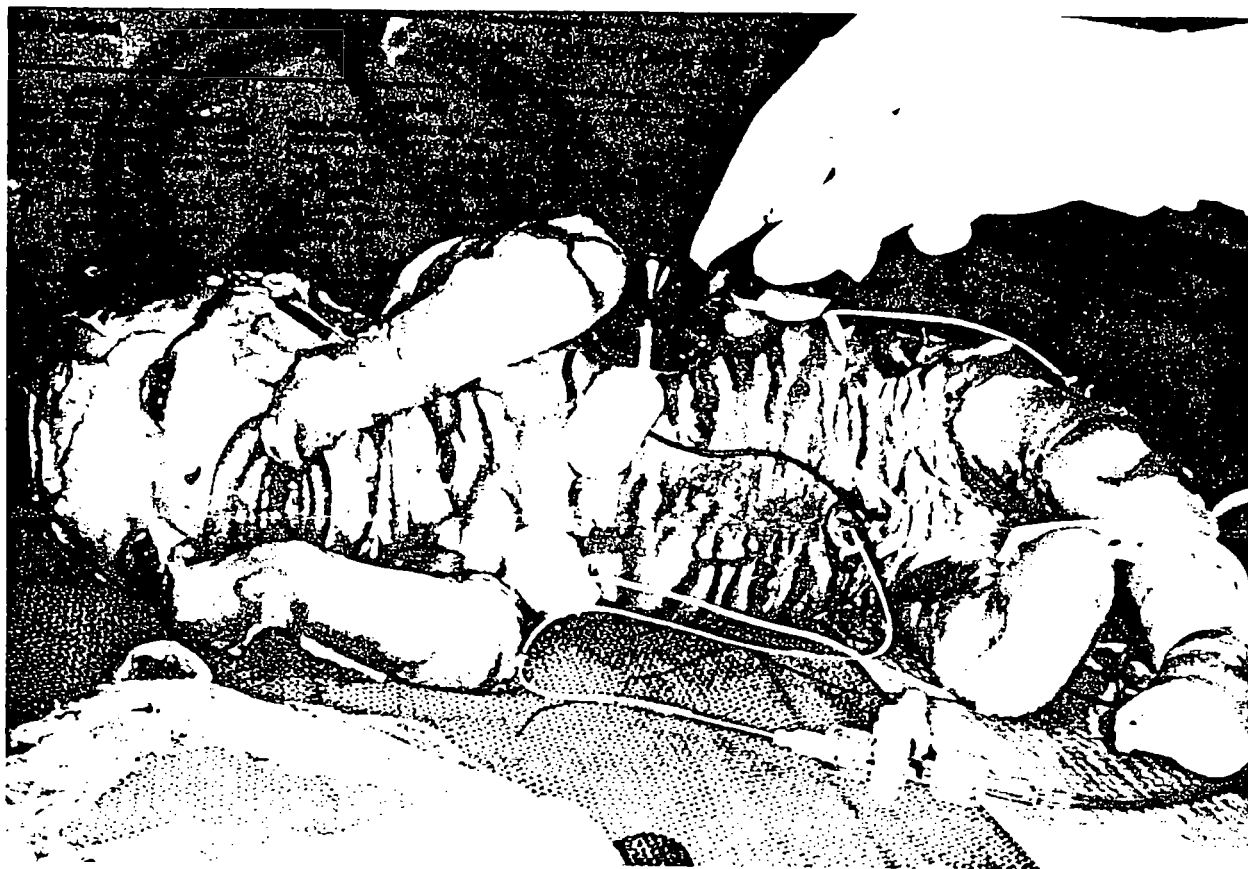


Fig. 1. Patient no. 1, a harlequin fetus who died on the first day of life.

ectropion of each eye and poorly developed nose and ears. Tightness of the skin of the hands and feet led to digital deformities. A piece of amnion was adherent to the skin. In the areas of fissuring, for instance, over the abdomen, redness was visible between the areas of the hyperkeratosis. Over the buttocks and the feet large plaques formed a hard, waxy coat.

The infant was covered with Saran wrap, and several warmers were used. A rectal probe was inserted, and treatment was initiated with dextrose and water, ampicillin, gentamicin, and morphine. Sterile technique was employed. The leukocyte count was 16.9/cm, with 23% polymorphonuclear cells, 2% bands, 71% lymphocytes, and 4% monocytes. The hemoglobin and hematocrit were normal. The infant's partial pressure of carbon dioxide rose, and on the evening of admission her heart beat and respiration ceased.

At autopsy the major pathology was in the skin and lungs. The infant had quite marked thymic hyperplasia, and her spleen was large. There was a moderate degree of pulmonary hypoplasia. Adjacent to the cracks was acute inflammation and edema.

Patient No. 2

This boy was born at Kaiser Hospital, San Diego, weighing 2270 g (<5th centile) and measuring 46 cm long (5th centile). The mother was a 24-year-old, gravida 1 para 0, and the pregnancy had been uncomplicated until spontaneous rupture of the membranes at 33 weeks of gestation. Labor began 15 days later, and delivery was precipitous. The umbilical cord had been avulsed approximately 5 cm from the infant's body. It was promptly clamped. The characteristic features of harlequin ichthyosis were apparent immediately, but the diagnosis was not made before an endotracheal tube was placed and assisted ventilation initiated. The parents were informed of the usual prognosis and requested treatment. By day 5 the ventilator was discontinued. Electrolytes were carefully monitored, as were blood urea nitrogen and creatinine, and there were no abnormalities. Electrolytes were measured daily for the first 8 days and frequently over the next week. The infant was given generous amounts of fluid because of experience with collodion babies, but there never was a problem. On

day 8 the sodium was 143 and potassium 3.5 mEq/L. He was initially treated in an isolette in a high-humidity atmosphere. The infant remained in the special care nursery for the first four months of life.

By 16 days of age his appearance (Fig. 2), originally very similar to that of patient no. 1, had improved. The thick surface scales could be seen on roentgenograms of the chest. The hands and feet at birth were both very swollen, and the fingers were webbed. The ears were adherent to the scalp.

Therapy with retinoic acid (Accutane) 1 mg/kg/day was begun on day 5. By day 13 necrosis of the tips of some toes was evident. With time, the large, thick scales gave way to fine, flaking scales. The black necrotic tips came off the fingers. The ears remained adherent to the scalp. By 37 days the skin was very red and shiny. On treatment day 47 retinoic acid was stopped, and 6 days later the skin was somewhat more cracked and flaking. The perianal or diaper area



Fig. 2. Patient no. 2, a harlequin fetus at 16 days of age. Treatment with retinoic acid had been initiated.

always appeared better than the rest of the skin. The skin in all areas healed very quickly. A circumcision was required because of phimosis and urinary retention, and it healed completely within two days. A punch biopsy in the buttocks area at 70 days of age healed in 2 days. This unusually rapid healing of biopsied skin was documented photographically.

Despite strict reverse isolation, the infant had problems from early on with infections of the skin, which appeared unusually susceptible to local infection. Superficial pustular skin infections, infections under the scale, and infections under the nails were continuing and recalcitrant. A variety of infecting organisms was identified, including *Staphylococcus aureus*, *Staphylococcus epidermidis*, and *Pseudomonas aeruginosa*. The patient had a good systemic leukocyte response. His infections were treated locally and systematically with antibiotics, but he virtually always had some pustules.

Because of very tiny ear canals, examining his ears was difficult and required an otolaryngologist using an operating microscope. The infant had a number of ear infections starting at 3 months of age. On computerized tomographic (CT) scan the ossicles of the middle ears were large, and there appeared to be fusion between the malleus and incus. Fluid was demonstrated in the mastoids on a number of occasions. Hearing was diminished, as demonstrated by repeated brain stem auditory evoked responses. Both CT and magnetic resonance imaging scans and ultrasound of the head revealed dilated ventricles. The appearance was stable. A small left ventricular hemorrhage in the neonatal period was documented by ultrasound without clinical signs.

Despite nasogastric alimentation from age 3 weeks, the baby failed to thrive and to develop. His extremely large fontanelle had not closed by 15 months. By this time he had developed six teeth. The ectropion was reversed, but he continued to have trouble with his lids and required lubricating drops at night. His skin continued to be bright red with many dry scales. His hair was sparse, thin, and blond. His toenails were thickened and often misshapen, and some were missing. The patient was intolerant to heat; he did not sweat well and readily became febrile. Air conditioning was effective in managing his heat intolerance.

At 15 months of age he was referred for admission to the Clinical Research Center of the University of California San Diego because of failure to thrive. At this time he displayed virtually complete failure to grow. His weight was 4 kg (50th centile for about 3

wks of age), height 60 cm (50th centile for about 11 wks) (Fig. 3), and head circumference 44 cm (50th centile for 6 mos) (Fig. 4). He was not developing; he was not even sitting or holding his head with security. His developmental age (DA) was assessed as 2 months or less at chronologic age (CA) 15 months.

Nutritional history indicated that this boy was taking a large number of calories. A count of the mother's dietary recall indicated 296 calories/kg/day from a combination of a high-calorie formula and pureed foods.

On admission, nitrogen balance studies were carried out while the infant was fed his usual diet. He ingested 227 calories/kg/day and 4.2 g/kg/day protein (Table 1). He put out very little nitrogen in urine or stool. An attempt was made to collect all of the desquamated skin during this period, but this proved not to be possible. Analysis of the nitrogen content of the material collected indicated it to be essentially

pure protein. The grams of protein calculated from the nitrogen content equaled the grams analyzed. On the basis of these findings, it was decided that the turnover of protein to make keratin was so rapid that the child required an additional 2 g/day nitrogen to meet his protein requirement. These calculations reflected a 2-g apparent positive balance despite the fact that the patient was not growing. Dietary inadequacy of protein was substantiated by the serum concentration of albumin of 2.9 g/dl.

It was also evident that it was not physically possible for him to eat any more than he already was. The initial approach was to insert a nasoduodenal feeding tube and to institute continuous overnight feeding, while he continued with his usual diet during the day. This regimen provided a minimum of 8 g/kg protein/day and 220 calories/kg. A special formula was developed that contained 500 ml Enfamil concentrate with iron, 29 g Rapidly Dispersing Protein powder, and 100 ml water (24-hour total volume 600 ml). The formula contained 1.3 kcal/ml and 37.5 g protein. Additional supplementation included 0.4 mg cupric sulfate, 0.04 mg chromic chloride, 40 mg magnesium sulfate, 5 mg zinc acetate, 50 µg folic acid, 400 IU vitamin D, 70 mg vitamin C, and 6 ml ferrous sulfate. The patient was fed his usual pureed foods and formula ad lib during the day every two to three hours, and the remaining formula was given by tube at 40 ml/hour from approximately 9 P.M. to 9 A.M. while he was asleep.

The patient tolerated this feeding program well, gaining 690 g in 12 days. His growth response was an essentially vertical acceleration of weight. Growth in height followed in a less dramatic but predictable fashion. His head grew quite rapidly. He continued to grow at this rate following discharge. Dramatic results occurred both physically and developmentally. The pinnae became defined and less adherent to the scalp. By 24 months he could sit independently and support himself standing, and he had a few words; developmental age was estimated at 1 year.

By 2 years of age the weight curve began to flatten because the boy no longer tolerated the nighttime feeding tube; when it was inserted he simply pulled it out. Therefore a plan was developed to increase his protein intake during daytime feedings. Dietary intake excluding his nighttime feedings at this time was 120 calories/kg and 3.9 g/kg protein. Protein supplements were added to the regimen, and additional calories in the form of polycose and corn oil were used, together with added feedings during the night whenever he awakened. The dietary goals

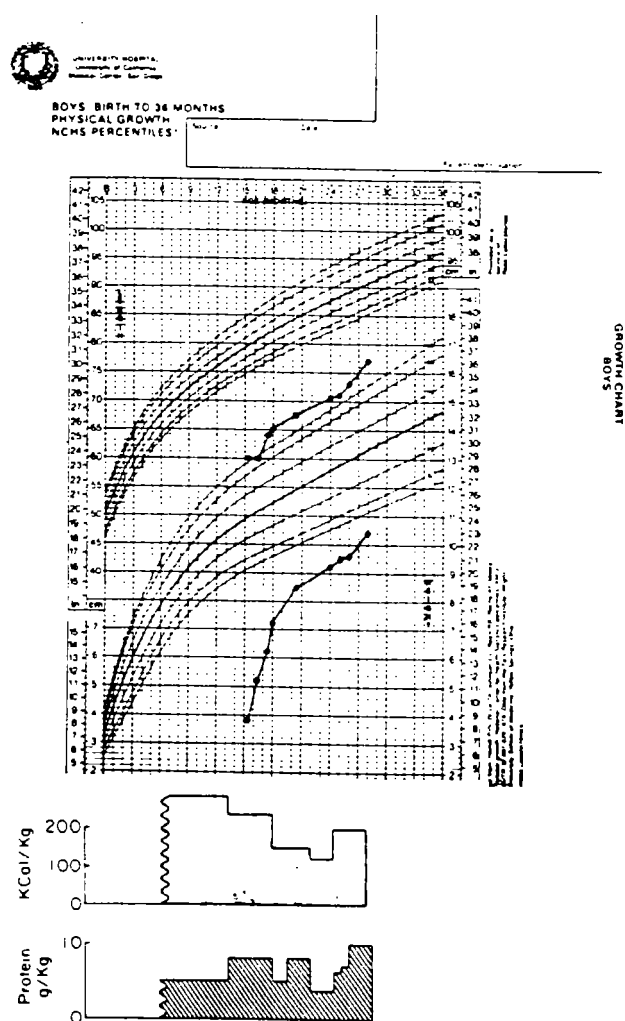


Fig. 3. Growth in height and weight of patient 2.

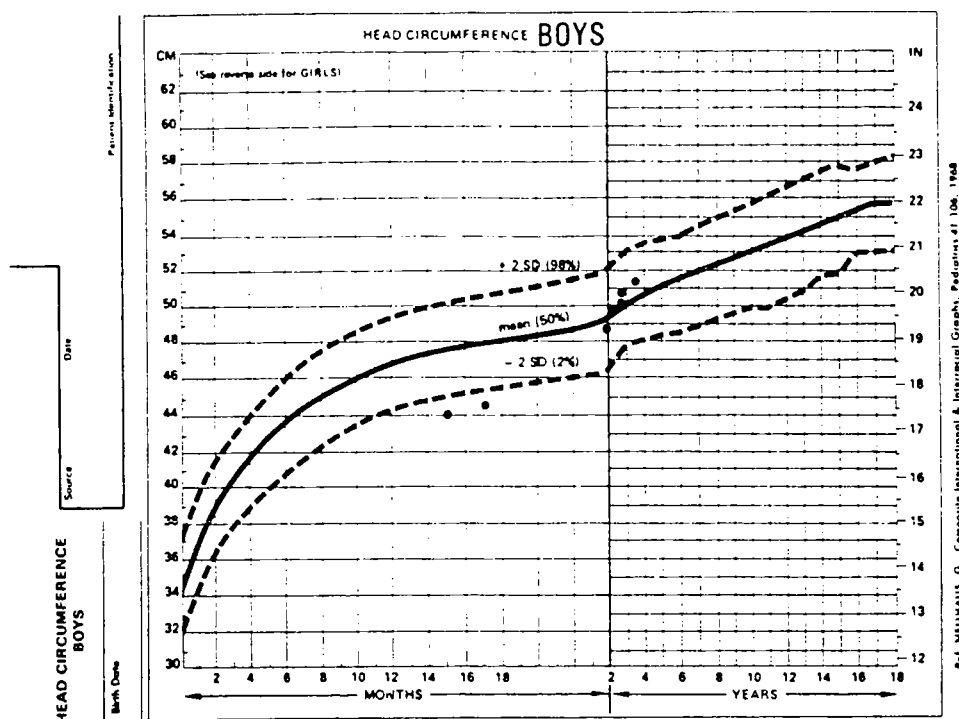


Fig. 4. Head circumference of patient 2.

were 200 calories/kg and 10 g/kg protein. With these changes growth resumed without tube feeding. Growth in weight and head circumference was striking. The concentration of serum albumin increased from 2.9 g/dl at 15 months of age to 4.1 g/dl at 28 months.

Developmental milestones improved as he grew. By 3 years of age the boy could walk rapidly with a broad-based gait, but could not run, and he spoke several words and phrases. By 4 years, he could run and spoke in sentences. By 5 years of age his speech content was judged appropriate for a

4-year-old. A formal Stanford Binet I.Q. test at CA 3.9 years showed him functioning at a MA of 2.6 years.

DISCUSSION

Both infants were typical examples of the harlequin fetus. Clinical characteristics included low birth weight and prematurity, the latter being more common than term birth. Dense hyperkeratotic scale and deep fissures produce the characteristic appearance of the skin. Contracture of the thickened skin leads to

Table 1. Nitrogen Balance and Serum Albumin Levels

Day	Age (mos)	Nitrogen (g)					Serum Albumin (g/dl)
		Intake	Excretion		Total	Balance	
			Urine	Stool			
1	15	2.85	0.07	0.15	0.22	+2.63	2.9
2	15	2.99	0.30	0.15	0.45	+2.54	
6	15	4.70	0.51	0.83	1.34	+3.36	
7	15	6.48	0.36	0.83	1.34	+5.29	
395	28						4.1

The apparent positive nitrogen balances on days 1 and 2 are considered to approximate losses in the skin.

ectropion and similar changes about the mouth (eclabion). Ears and nose may be adherent to the scalp or poorly defined because of fusion to the surrounding skin. There may be contractures of the digits or extremities; in our patient the tips of some toes became blackened and necrotic. Death in the neonatal period appears usually to be pulmonary in origin, as illustrated by the first patient. The tight skin of the chest is thought to produce constrictive forces that lead to pulmonary hypoplasia and restriction of postnatal respiration. In patient no. 1 the lungs were hypoplastic as well as diminished in size; despite this the baby had good initial gas exchange. The increasing carbon dioxide thereafter appeared to reflect a restrictive respiratory problem as the skin dried around the chest. Experience with assisted ventilation for five days in patient no. 2 suggests that this problem may be of short duration. It is also possible that some infants do not survive because the prognosis is viewed as hopeless and no treatment is given during this initial period.

Survival of the neonatal period in a harlequin fetus was first reported in a Bangladeshi girl in England.⁴ She was 2½ years of age at the most recent report.⁵ She is now lost to follow-up. She was treated with supportive care in the neonatal period and also was given etretinate. The rationale came from the experience with ichthyosis and psoriasis in which the retinoid decreases the size of desmosomes in the keratinocytes of hyperkeratotic skin.³ It may also be relevant that deficiency of vitamin A may lead to hyperkeratinization. It was for this reason that treatment with retinoic acid was begun in our patient no. 2. Etretinate was not available in the United States. Another harlequin fetus who survived more than two years was reported from Australia⁶ and a 9-year-old was reported from Texas.⁷ Progression to ichthyosiform erythroderma was also observed in the latter. She was reported to have normal intelligence and to excel in school.

Nutritional aspects of dermatologic disease in infancy have not been emphasized. Failure to thrive was also prominent in the patient reported from England, who, like our patient 2, was well below the 3rd centile in growth and had appreciable delay in motor development. Experience with our surviving patient indicated that the enormous loss of desquamated skin produces a requirement for protein and calories that may be impossible for even a very hungry infant to meet. Initially, we overcame this problem by overnight nasoduodenal feeding of a high-protein formulation. Recognition that the loss was virtually pure

protein led to a later regimen sufficiently high in protein to promote growth and development without supplemental tube feeding.

The conclusion that protein had been limiting was drawn from our experience on two occasions with this patient when change in protein intake without alteration in total calorie intake led to enhanced growth. This was supported by the data on nitrogen balance and the serum concentration of albumin, which in this patient was also a marker for protein adequacy as indicated by an increase from 2.9 to 4.1 g/dl with the provision of increased amounts of dietary protein. The amounts of protein required were enormous, amounting to 8 to 10 g/kg, and similar to those seen in children with severe burn injury. Caloric requirements were also excessive, ranging from 200 to 296 kcal/kg. It was clear that the patient had been capable of a high-calorie intake of this order of magnitude prior to study. The change in regimen that led to growth and development was the increase in the intake of protein. Once protein needs were met, adequate growth and development followed. The rapid catch-up growth in head circumference and subsequent development are consistent with the concept that the synthesis of proteins of brain cells is an absolute requirement for development to proceed.

The histopathology of the harlequin fetus is that of a congenital ichthyosis in which the light microscopic picture is that of marked hyperkeratosis.⁹ Electron microscopy reveals very large desmosomes, thickened stratum corneum with lipid inclusions, and absent or abnormal lamellar granules.^{3,10} Study of epidermal structural proteins¹¹ revealed heterogeneity in 10 harlequin fetuses, one of whom was our patient 2. Three phenotypes were recognized. In type 1, represented by patient 2, profilaggrin was not converted to filaggrin. Survival of the neonatal period permitted observation of an evolution of the dermatologic picture to that of ichthyosiform erythroderma.⁵ This form of ichthyosis is transmitted in an autosomal recessive fashion, consistent with what is known about the genetic transmission of the harlequin fetus. In the English report⁴ the surviving infant was their fifth harlequin fetus. The other four died. In another family¹¹ four harlequin fetuses, three males and a female, were born to consanguineous Spanish parents. It is not clear whether the harlequin fetus does or does not result from the same defective gene as other forms of nonbullous ichthyosiform erythroderma.^{5,12-14} Similar appearance, histopathology, and pattern of inheritance do not establish that they are not distinct diseases. In fact, most patients with

congenital ichthyosiform erythroderma do not have the extreme phenotype of the harlequin ichthyotic patients.

The harlequin ichthyosis may be confused with the other neonatal ichthyosis, the collodion infant.^{14,15} In the latter, a thin type of almost cellophane or collodion membrane surrounds the infant. There is ectropion of the eyes and some fissuring around the mouth, but the skin is not as tight as it is in the harlequin fetus. There is not much fissuring. Most collodion babies survive with treatment, whereas most harlequin fetuses are not aggressively treated. The collodion infants may have problems of neonatal fluid and electrolyte management; care must be exerted to avoid massive dehydration and hypernatremia, which may occur very quickly. Neither dehydration nor electrolyte imbalance was observed in patient no. 2, but it is recognized that excessive water loss from the skin is a potential problem in any infant with an ichthyotic dermatosis.¹⁵ Both harlequin and collodion infants may have problems maintaining body temperature in the neonatal period and handling heat stress thereafter. Air conditioning appears to be sufficient for management of heat intolerance. Skin infections are a problem in both types of severe ichthyosis. Pustular disease of the skin remained a continuing problem in our patient.

SUMMARY

Survival is possible in some harlequin fetuses. They require extensive intensive care with careful attention to respiratory status, and electrolyte and fluid requirements. Our surviving patient had failure to thrive and developmental delay that were markedly improved with correction of a severe protein deficiency. The exact cause of the deficiency is not completely worked out but is most likely directly related to the large loss of desquamated skin in these patients. Continued monitoring of growth, development, and nutritional requirements is critical in surviving children.

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Harlequin Baby Treated with Etretinate

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Abstract: A child born with the harlequin fetus abnormality survived and showed a surprising degree of spontaneous improvement in the first six weeks of life. At this age etretinate was instituted, producing considerable further resolution. The child is now over 2 years old.

In 1985 a patient was described with the harlequin fetus abnormality who survived with intensive neonatal care, and in whom etretinate was used from an early age (1). The authors made the point that these infants do have a potential for survival, and that etretinate may produce considerable improvement in the skin condition. Encouraged by their report, we elected to use this drug to treat a patient who had survived for six weeks.

CASE REPORT

The patient, a male, was the fifth child of consanguineous Lebanese parents. One sibling was alive and well and one had the Richner-Hanhart syndrome. Two others had been harlequin fetuses (Figs. 1 and 2) and died a few days after birth. Our patient was born at term by emergency cesarean section due to breech presentation and fetal distress. Birth weight was 3220 g, just below the fifth percentile. At birth his appearance was similar to that of his two deceased siblings. Out of consideration for his parents' photographs were not taken. The infant was covered with a tight, shiny, whitish membrane scored by deep red fissures. The fingers and toes were held in a position of flexion by tight, thick skin, and the tip of one digit was cyanotic. Nails were present on all digits. He had ectropion of both upper and lower eyelids with marked conjunc-

tival chemosis. There was eclabium, with the mouth held open. The nares were patent. The ears were deformed, being folded and tethered to the scalp. Eyebrows and eyelashes were absent, but quite profuse hair grew through the thick plates of scale that covered the scalp. General physical examination was otherwise normal.

The child was not expected to live but was given supportive treatment, being nursed in a humidifier (isolette) and fed regularly by tube and bottle. He had three baths a day in warm water with oil added. Artificial tears were instilled into his eyes, and analgesia with perhidine was given as required.

Over the next few days the membrane dried and cracked into thick, yellow, crusted plates. The digits became more mobile as the membrane cracked and peeled off. The tip of the left fifth finger, which had shown cyanosis, became necrotic and was lost, but the other digits remained intact.

In the next three weeks some of the plates of scale separated, while others, some of which became very dark, remained around the head, buttocks, and lower legs (Figs. 3-5). Elsewhere there was a coarse, lamellar scale. Apart from the areas of dark scale, the whole skin had a yellowish color. He had deep fissuring in the major flexures with secondary monilial infection. The ectropion and eclabium improved and the ears unfolded to some extent.

At 3 weeks of age the patient developed a fever.

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Figure 1. First harlequin fetus sibling.

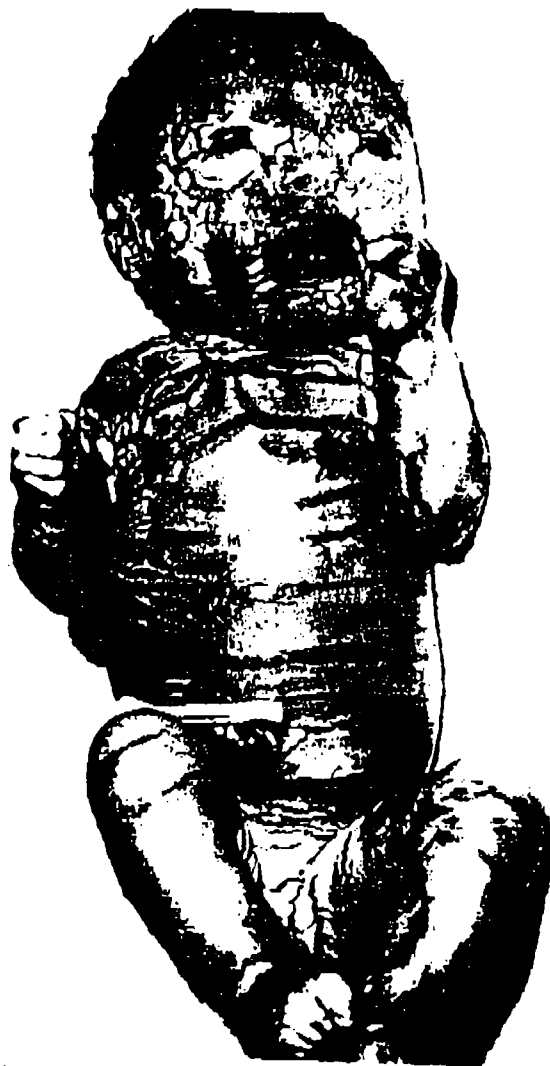


Figure 2. Second harlequin fetus sibling.

and *Staphylococcus aureus* was grown from skin, conjunctivae, and blood. He was treated with intravenous flucloxacillin and gentamycin, and responded well.

At age 6 weeks he was clearly going to survive, and after careful discussion with his parents, who were anxious for active intervention, a decision was made to institute retinoid therapy.

Screening tests performed at this time showed a hemoglobin of 7.9 g per dl, white cell count of 11.8×10^9 per L, with a normal differential and a platelet count of 413×10^9 per L. Results of liver function tests and fasting levels of cholesterol and triglycerides were normal.

Etretinate was commenced in a dosage of 4 mg per day, just over 1 mg per kg. Eucerin ointment was used as an emollient, and clioquinol 0.5% cream was applied to the flexural areas that showed secondary monilial infection. After three weeks of treatment the skin had thinned dramatically over most of the body (Fig. 6) and was now erythema-

tous rather than yellow. The large plates of scale had been lost from all areas except the face and scalp. The fingers and toes became fully mobile and the ears unfolded further. The ectropion was now minimal, although the lids remained tight. Over the next few weeks all the thick plates of scale resolved.

At age 14 weeks, osteomyelitis of the left hip developed with dislocation of the left femoral head. This was treated with traction and intravenous flucloxacillin followed by oral chloramphenicol.

Over the first four months of life the child failed to thrive. His weight fell to well below the third percentile despite the institution of a high-calorie diet. His height also was well below the third percentile.

Anemia was a persistent problem and was thought to be due to chronic infection. He also de-

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Figure 3. The patient at 4 weeks of age, prior to etretinate therapy, has a thick scale on the face, entropion, and eclabium.

veloped an acute episode of thrombocytopenia for which no cause could be found other than infection. He was given a transfusion of platelets and subsequently, whole blood.

Developmental assessment at 6 months showed



Figure 4. Plates of scale on the scalp (age 4 wks).



Figure 5. Plates of scale on the buttocks (age 4 wks).

that he had good head control but could not sit out support. He was alert and was responsive to his parents. His skin continued to improve. He was discharged from the hospital at age 6 weeks.

At 1 year of age his height and weight increased to just below the third percentile. Mentally, he had progressed significantly, assessed as normal in intellectual and fine motor functions, but still delayed by three months in gross motor milestones. His dosage of etretinate per day now was equivalent to 0.5 mg/kg. His skin maintained its improvement. A bone technetium scan performed to detect signs of noid toxicity was normal, and results of liver function tests and lipid levels remained within limits.

At the most recent assessment at 2 years of age, his height was still below the third percentile, weight was in the tenth percentile. He was approximately six months delayed in language and fine motor skills, and his gross motor development was further delayed by his damaged hip. He was cheerful, and well integrated into his family.



Figure 6. After three weeks of etretinate therapy the patient shows considerable improvement.

skin remained red and scaly, clinically resembling a congenital ichthyosiform erythroderma (Fig. 8).

DISCUSSION

Since the original description of a harlequin fetus in 1750 by the Reverend Oliver Hart in his diary, over



Figure 8. The patient at 2 years of age.

100 cases have been described in the literature. These have come from many countries, with reports in the last few years of cases from India (2-5), United States (6-8), Greece (9), South Africa (10), Libya (11), Spain (12), and the United Kingdom (in a Bangladeshi patient) (1). The condition appears to have no racial predilection. Our patient was born in Australia of Lebanese parents.

The condition was recently reported in dizygotic twins (8), and several patients have had a history of one or more involved siblings (1,10,12). It occurs equally in males and females. Parental consanguinity was present in several cases (2-4,8,12). The disorder has not been reported in successive generations. The features suggest that it is determined by an autosomal recessive gene.

Our patient was the third harlequin fetus born to his consanguineous parents. Another sibling had the Richner-Hanhart syndrome, also an autosomal recessive condition.

The exact nature of the harlequin fetus abnormality is unclear. It has been suggested that it may represent the most severe form of nonbullous congeni-



Figure 7. The patient at 1 year of age.

tal ichthyosiform erythroderma/lamellar ichthyosis (13). It was pointed out, however, that congenital ichthyosiform erythroderma and harlequin fetus have not as yet been reported in the same family (14). Another possibility is that it is a separate form of autosomal recessive ichthyosis. Finally, it may represent a phenotypic expression of several genotypes akin to the situation with a collodion baby (15). Evidence in support of this last hypothesis is the finding in some cases, but not others, of abnormalities in stratum corneum keratin (6) and epidermal lipids (7).

The clinical features of harlequin fetus reported in the literature are strikingly similar. The whole body is covered with a thick, encasing membrane that is often white at birth but soon becomes brown, gray, or yellow. It is divided into segments by deep bleeding fissures. Severe ectropion is present, usually with marked conjunctival chemosis, and eclabium with the mouth held open in an O shape. Hands and feet are often edematous, with the fingers and toes held in a flexed position producing a clawlike deformity. The encasing membrane may compromise the circulation, leading to loss of parts of the digits. Larger joints may also be held in a position of flexion by the membrane. The ears are folded and distorted. Eyebrows and eyelashes are often absent and scalp hair may be sparse or absent. In patients who survived for a few weeks, the membrane peeled off in some areas but soon reformed, producing large, dark, hyperkeratotic crusts.

Our patient had a clinical picture very similar to that of the other reported cases. By 6 weeks of age, before etretinate was commenced, he had had a significant degree of improvement, with a partial reversal of ectropion, eclabium, ear folding, and flexion deformities of the digits.

Other abnormalities reported in these infants include nonspecific renal tubular abnormalities, thymic aplasia or hyperplasia, thyroid aplasia, cervical hemivertebra, and hypoplasia of the lung (3,10,11). In our patient no abnormalities in other organ systems were detected at birth. His course was complicated at a later stage by septicemia and osteomyelitis, and his height and weight have remained below the third percentile.

Most patients have died in the first days or weeks of life, with death suspected to be due singly or in combination to septicemia, pulmonary infections, mechanical restriction of breathing, and disordered temperature regulation. Many of these infants were born prematurely, which almost certainly contributed to their early death.

One patient survived to age 9 months before dying from sudden infant death syndrome. The survival was attributed to intensive care in the first weeks in an incubator and the early use of parenteral antibiotics (7). In the first reported use of retinoids in this condition, etretinate was begun at 2 days of age (1). In addition, the child was nursed in an incubator, adequate fluid and caloric intake was assured by the use of a nasogastric tube, parenteral antibiotics were given, and anemia was treated by transfusion and iron supplementation. The author implied that intensive care, rather than the retinoids, was responsible for the child's survival.

Lawlor (15) communicated with three other dermatologists who are caring for surviving harlequin fetuses treated with intensive care and retinoids. It is clear that these infants do indeed have a potential for survival, with the chance enhanced by early, intensive nursing care and possibly the use of retinoids. Our patient showed some resolution prior to the use of retinoid, but dramatic improvement was obvious within weeks of commencing etretinate in a dosage of just under 1 mg per kg per day; this is maintained at a dosage that now represents approximately 0.5 mg per kg per day. In Lawlor's patient etretinate was ceased at 14 months of age with only minor worsening of the condition. That patient is now over 3 years old and is progressing well (F. Lawlor, personal communication, 1988). We intend to withdraw etretinate in several months to assess the continuing requirement for the drug. As yet no signs of etretinate toxicity have occurred.

CONCLUSION

Now that there are several surviving harlequin fetuses, it is suggested that the term "harlequin baby" may be more appropriate (15). It is clear that these babies may survive, and if they do, etretinate can significantly ameliorate the skin condition. Early, intensive nursing care will greatly enhance the potential for survival. Because of the probable severe lifelong ichthyosis, even with treatment, a dilemma remains as to how intensive early treatment should be.

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Etretinate Treatment For Harlequin Baby

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ABSTRACT

This report of a Harlequin foetus surviving is due essentially to two factors.

- a) *the intensive care provided to the erythrodermic infant, looking after its nutrition, temperature control and controls of infection and*
- b) *the use of a new synthetic Vitamin A acid - Etretinate in the management of dyskeratinisation syndromes.*

As this is of autosomal recessive inheritance, besides genetic counselling, Singapore has to develop its prenatal diagnostic techniques viz - amniocentesis and ultrasound studies of the skin, to detect dermatological problems.

Key words : Harlequin, ichthyosis, erythroderma, ultrastructural studies, etretinate

CASE REPORT

A Chinese female infant was born via caesarian section at 37 weeks gestation, because of a difficult breech presentation, at Kandang Kerbau Hospital in 1991. She is the first born of non consanguineous parents. The birth weight was 2250 gm and APGAR score was 9 at birth. At birth the entire body was encased by a thick keratinous casing which fissured irregularly, exposing underlying erythroderma, looking like a Harlequin foetus.

The face showed severe ectropion and eclabium, with absent hair, eyebrows and eyelashes. The nasal bridge was flattened. She had a taut "fish mouth", folded ears which encased by sebum and the auditory canal were occluded by keratin. The hands and feet were encased in keratin, causing ainhum, and subsequently dry black gangrene.

MANAGEMENT

The baby was nursed in a humidicrib in the neonatal intensive care. She was monitored closely regarding nutrition, treatment of any sepsis and its prevention, with good temperature control and care of the exposed

cornea and skin management. She survived the first month, being tubefed and had oil baths daily with careful removal of thickened skin. The skin culture did not yield any bacterial growth. However, bacitracin ung, 2% (Mupirocin) was applied to fissures and prophylactic cloxacillin, crystalline penicillin and gentamycin were administered. Neosporin eye drops were instilled and intravenous ceftazidime given when the latter culture from the eye grew *Acinetobacter*. Corneal drying was prevented by artificial tears and eye pads. A skin biopsy confirmed ichthyosis, showing hyperkeratosis and granulosis and irregular acanthosis.

The decision to give etretinate was made only after the infant had reached a stable state at 6 weeks of age, at the dose of 1 mg/kg body weight, ie. 2.5 mg/day. Within a fortnight, dramatic improvement was seen with remarkable shedding of the thick scales, and new scales produced were papery thin. Ectropion remarkably improved. The movement of the four limbs were increased. Ammonium lactate 12% (lacydrin) applied regularly was added to the skin. At six months, the dose is 5 mg/day, 1 mg/kg body weight.



Fig 1: Harlequin foetus at birth with diamond shaped scale

Though small at the third percentile for weight and height, she has normal developmental milestones, and appears unimpaired intellectually.

The initial screening profile was normal. At 6 months of age, her haemoglobin is 10.2 g/dl, liver function test is normal, S. albumin 42 g/l (36-50), protein 62 g/l (65-80), alkaline phosphate 259 U/L (38-126). Normal results are in parentheses. Serum transaminases, triglycerides and cholesterol are normal.

DISCUSSION

The place of the Harlequin foetus in the classification of ichthyosis is controversial and the precise mechanism of disordered epidermal carnification is unsure. Clinically, it is like a nonbullous congenital ichthyosiform erythroderma.

The ultrastructural studies of the harlequin fetal skin show large concentric lamellar bodies with focal absence of intercellular lipids in the stratum corneum, with giant desmosomes and tight desmosomal contracts. The loss of lipids result in build up of the giant lamellar bodies. All these contribute to a retention mechanism of ichthyosis⁽³⁾.

INTRODUCTION

The harlequin foetus was first described by Hart (1750). Since then, most of these infants perish as a result of systemic complications like sepsis from deep skin fissuring, respiratory problems, pneumonia and



Fig 2: Baby at 9 months, after etretinate treatment

feeding difficulties, poor temperature control or prematurity.

Both excellent neonatal intensive care and oral retinoids, Etretinate, has drastically benefited and changed the prognosis of these infants, and allowed long term survival and improved quality of life.

Retinoids are shown to produce changes in both the tonofilaments and reduce the size of desmosome; resulting in improved regulation of epidermal proliferation⁽⁴⁾. Isotretinoin, etretinate and acitretin have shown good result in the treatment of congenital ichthyosis.

In the growing child, osseous development has to be monitored closely, to see if growth is stunted. The child is being monitored for all the side effects of etretinate (5-10).

In our management of two children with congenital ichthyosis on etretinate cases over a decade, radiological studies of bones yearly has not shown any growth retardation, cysts or hyperostosis. The liver function and serum lipids have not increased. Muco cutaneous symptoms like lip chapping is the chief complaint. Curly hair is a welcomed side effect in these children. The intermittent withdrawal of retinoids leads to rapid deterioration, with severe ichthyosis. This is socially unacceptable to the parents and our preferred therapy is alternate day therapy at the minimum dose. The etretinate will be stopped as the children

improved. Our experience has supported the data that children may tolerate a higher dose of etretinate than adults.

Our experience with etretinate in the Harlequin foetus has been very encouraging and appears to give a marked aesthetic difference to this severe form of ichthyosis.

However, this is possible as the infant may be a milder clinical phenotype and can sustain respiration. Improved neonatal intensive care, adequate nutrition, prevention of sepsis has allowed the child to retreat with etretinate.

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Management and follow-up of harlequin siblings

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Summary

Harlequin fetus is a rare clinical entity, and survival of affected infants beyond the first year of life is uncommon. Management involves intensive care of the skin and eyes, close monitoring of fluid and electrolyte status, constant support and counselling of parents, and surveillance against infection and side-effects of medication. A well-coordinated multidisciplinary approach can prolong survival beyond the neonatal period.

We report our experiences in the management and follow-up of two successive harlequin siblings.

'Harlequin fetus' is the descriptive term for the most severe form of congenital ichthyosis. The earliest record of its description is from the diary of the Reverend O.Hart in 1750.¹ Since then it has been referred to as 'fetal ichthyosis', 'ichthyosis intrauterina', 'keratosis diffusa fetalis', 'congenital diffuse maligna keratoma', 'malignant keratosis' and 'alligator baby'.

Case reports

Case 1

A male infant was born by assisted breech delivery after spontaneous onset of labour at 37 weeks' gestation, to a gravida II, para I, 23-year-old Saudi woman. The antenatal period had been uneventful. The baby had good apgar scores, and the birth weight was 2460 g. The parents were first cousins. A 3-year-old sibling was in good health.

The clinical appearance of the baby was striking (Fig. 1). The skin was hard, thickened, waxy and yellowish in colour. It was split irregularly to reveal erythematous, moist fissures. The limbs were in a semiflexed position, and had limited mobility. The fingers and toes were hypoplastic. There was severe ectropion and eclabium. The nose was deformed, and the ears were rudimentary. The baby's cry was normal, but he was unable to suck effectively.

Immediately after transfer to our neonatal intensive care unit, the baby was nursed in a humidified incubator. As peripheral venous access was difficult, an umbilical venous line was set up. For the first few days this route was used as intravenous access. An extra 25% allowance was provided for fluid and calorie requirements from day 1. After taking appropriate cultures, antibiotics were commenced.

Care of the skin consisted of 2-hourly applications of paraffin ointment (a mixture of equal parts of white soft paraffin and liquid paraffin) to the whole body. Areas which had tight bands were covered with topical 10% urea in liquid paraffin, applied under occlusive dressings which were changed every 6 h. The baby was bathed twice daily using Alpha Keri® bath oil (Bristol Myers).

The eyes were kept moist at all times with methylcellulose drops and ointment. The plugs of keratin in the auditory canals were cleared with Cerumol® drops. Histology of a skin biopsy specimen taken on day 3 revealed marked hyperkeratosis, mild parakeratosis and prominent rete pegs. There was a mild infiltrate of mononuclear cells in the upper and mid-dermis. Skin appendages were normal, and there were no bullae.

Oral etretinate in a dose of 1 mg t.d.s. was commenced. Full blood count, electrolytes, liver function



Figure 1. Case 1 soon after birth.



Figure 2. Case 1. Marked improvement—day 51.

tests, blood cultures, and culture of skin swabs were performed, and subsequently monitored regularly.

The skin gradually softened, and the hard yellow plaques separated, revealing an erythematous, scaly surface beneath (Fig. 2). Improvement in the skin around the mouth allowed the infant to suck from the bottle by day 4, and breast-feed by the age of 1 week. The range of movement in all the limbs improved, and the fingers and toes began to assume a more normal shape. The shape of the nose improved, but the external ears did not show any significant change.

As the skin improved, the frequency and intensity of topical treatment was reduced, and the baby was discharged home at the age of 51 days, weighing 3000 g. He was initially followed-up on a weekly basis.

During the next 21 months of follow-up, his general condition was stable. The parents coped well with his skin care, which was gradually tapered to application of paraffin ointment twice daily. However, his weight gain was poor; it remained below the 3rd percentile. At the

age of 18 months, he was alert and interested in his surroundings, and was able to make cooing and babbling sounds. His hearing and vision were normal. He had good neck control, but could not crawl, or sit up unaided. In addition to a few minor illnesses, he had three short-stay admissions to hospital for bronchiolitis, suspected septicaemia, and bronchopneumonia, at the age of 4, 9, and 15 months, respectively. Several other admissions were for respite care as his mother underwent cardiac surgery. At the age of 1 year, oral etretinate was discontinued.

At the age of 22 months he was admitted because of diarrhoea, vomiting and lethargy of 2 days' duration. His temperature was 37.2°C and the skin looked dry and neglected. Our aims were to rehydrate him, provide skin care, and investigate possible sites of sepsis, and these measures were promptly initiated. However, 24 h after admission he had a cardiorespiratory arrest, and did not respond to resuscitatory measures. Blood cultures grew *Staphylococcus aureus*. A post-mortem examination could not be conducted.

Case 2

A younger male sibling of case 1 was delivered vaginally at 38 weeks' gestation. He had good apgar scores, and his birth weight was 2560 g. His physical features were typical of harlequin fetus (Fig. 3). In comparison with his brother, the ectropion was more marked, but the fingers and toes were less hypoplastic.

The initial management was identical to that of the older sibling, including the dose of etretinate, and he was also treated with flucloxacillin and gentamicin. However, at the age of 2 weeks, when he was still in hospital, methicillin resistant *Staphylococcus aureus* (MRSA) was grown from the skin. He was isolated, and appropriate nursing precautions were taken. MRSA was not isolated from the blood. Hence, he did not receive parenteral antibiotics. Fortunately, the skin was clear of this organism within 2 weeks.

He tolerated oral feeds from the first day, started sucking from the bottle by day 4, and was breast-feeding by the end of the second week. His weight at the age of 5 weeks was 2665 g.

At five weeks of age his temperature began to fluctuate between 34.1 and 38.6°C. Abdominal distension and intolerance of feeds was noted. His skin, which had previously begun to improve, developed pustules and pockets of pus. There were areas of ischaemia and gangrene on bony prominences and the tips of the fingers and toes. At this stage *Pseudomonas aeruginosa*



Figure 3. Case 2 soon after birth.

was isolated from skin swabs. He was treated with appropriate antibiotics and supportive measures. Blood tests showed evidence of disseminated intravascular coagulation. He succumbed to this illness at the age of 44 days.

Discussion

Although there are several case reports of harlequin fetus in the literature,²⁻⁵ few affected infants have survived beyond the first year of life.^{4,6,7} Reports of subsequent siblings with the same disorder are infrequent.^{3,8} Ultrastructural studies of this condition indicate that harlequin ichthyosis is a genetically heterogeneous group of disorders, with altered lamellar granules and intercellular lipids, and variations in expression and/or processing of structural protein markers of normal epidermal keratinization.^{9,10}

Prenatal diagnosis by fetal skin biopsy is possible, and amniotic fluid cytology can be helpful.¹¹ The parents of

our patients were opposed to any intervention during pregnancy. Ultrasonic features of the harlequin fetus *in utero* have also been described.¹² Harlequin fetus may be associated with other congenital anomalies,¹³ but neither of our patients had any other congenital abnormality.

Both our patients were started on oral etretinate within the first few days of life. As in previously reported cases,^{3,4,7} we also noted a beneficial effect on the skin (Fig. 2). As the etretinate preparation we used was unstable, it had to be dispensed each day by the hospital pharmacy. However, dry diluent can be obtained (Avicel, 101, Honeywell & Stain Ltd, Wallington) to enable production of a stable powder for out-patient use.⁴ We monitored our patients closely for potential side-effects (including bone scans for skeletal abnormalities),^{14,15} but there was no evidence of any abnormality. In addition to medical treatment, intensive nursing care and dedicated care by parents is essential in the management of these babies.^{16,17}

We used paraffin ointment for topical therapy, and avoided the addition of salicylic acid, because salicylate toxicity in this setting has been described previously.⁴ During the period when we used topical application of 10% urea in liquid paraffin on areas with tight bands, we did not encounter any problems due to absorption of urea. Serum urea levels were all within normal limits (the highest recorded level was 6.9 mmol/l). However, once the bands had loosened, the use of this preparation was discontinued.

The development of digital gangrene, as occurred in our second patient, has been reported previously.² Septicaemia is the commonest cause of death in these babies, and death may be sudden,¹⁸ as was the case in our first patient.

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SUNDAY MAGAZINE

F SECTION ■ PITTSBURGH POST-GAZETTE ■ MAY 5, 1996



Eighteen-month-old Hunter Steinitz was born with an unusual skin disorder called Harlequin Ichthyosis. Hunter's skin grows so fast it cannot shed effectively. Here Hunter's mom, Patti, clears away loose skin in the outer ear during one of the two daily baths that Hunter gets to keep her skin supple.

The battle to trim medical spending could upset the network of care used to treat little Hunter Steinitz, who suffers from a rare skin disease

A life in the balance



Patti Steinitz recently visited with state Rep. Don Walko, D-North Side, to explain how the potential cuts in state and federal funding would affect children with disabilities. "Nobody can understand what we go through. I fight for her every day. ... I am always fighting someone."

The biggest worry Patti and Mark Steinitz had before their daughter's birth was what to name her.

Patti was 36 years old and felt good. Her morning sickness had abated early in her pregnancy. Sonograms and amniocentesis, the chromosomal abnormality test that also reveals the baby's sex, gave both mother and unborn daughter a clean bill of health.

They wanted a name that began with the letter H to remind them of Mark's late mother, Henrietta. They found the answer scrawled on a blackboard at a children's shoe store.

Hunter.

Story by **Ellen M. Perlmutter**

Photographs by **Annie O'Neill**

An unusual name for a girl, they admitted, but they liked it. It was Oct. 15, 1994, and Hunter Elizabeth Steinitz was due in five weeks.

That night, though, Patti's water broke. And she had no contractions.

"Something's wrong," Patti told her husband, as they dressed to go to the hospital. "Something has to be wrong."

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COVER STORY



Once a week, Hunter is visited by Sharon Clougherty, center, a child therapist, and Rebecca Klaw, right, a family therapist, both from ARC Allegheny.

Anne O'Neill/Post-Gazette photos

Hunter and her mom, Patti, visit with neighbor Rita Formhals. Patti feels that the love and constant stimulation at home have helped with Hunter's development.



Hunter's father, Mark Steintz, gives his energetic 18-month-old a dose of Propulsid, a drug that helps her stomach to empty more quickly. Her 17-pound body cannot afford to lose any nutrition.

Going to battle for Hunter

CONTINUED FROM PAGE F.1

On Oct. 17, after two days of bed rest at Magee-Womens Hospital, doctors induced labor.

Specialists arrived at the delivery room. They anticipated that the baby would require immediate attention for underdeveloped lungs, a common occurrence in preemies.

Hunter weighed 5 pounds, 3 ounces, and doctors immediately whisked her to a table across the room.

Mark, who had never experienced the birth of a child, thought nothing of the commotion. Patti was uneasy. She had an 11-year-old from a previous relationship, and the birth had been nothing like this.

Doctors put Hunter into an incubator and rolled it over to the delivery table where Patti still lay.

Hunter's skin was neon pink and covered with white, tough scales — like the hide of an elephant.

A doctor told the new parents: "Your baby has a skin disorder. It's called ichthyosis. We don't yet know what type, but we know it's severe."

The infant looked so raw that Patti wanted to know how much it hurt.

"She looked like nothing I'd ever seen before," Patti said. "She had bulging eyes. That's what I remember. Mark didn't know what to expect, but I knew. I knew. My baby wasn't well."

It seemed surreal for Patti, who grew up on the North Side and only two years before was a single mom. When she met Mark in June 1993 through friends, she was, and still is, a Pittsburgh police officer — first in Oakland, then on the South Side for 10 years before being transferred to the North Side station. Mark, who was living in Monroeville, had gone into his mother's picture-framing business with her, which he is now in the process of closing since her death. The

couple live in Observatory Hill on the North Side.

The condition that afflicted their daughter was rare. Few people have heard of Harlequin ichthyosis, also known as Harlequin fetus. A genetic disorder caused by the mating of two rare, recessive genes carried by both parents, the condition is present in just 1 to 5 of every 5 million babies born in the United States each year.

Hunter's skin grows so fast it cannot shed effectively. It thickens easily becoming scaly. Her body has trouble regulating its temperature. Dehydration can occur. The extra skin can tighten across her abdomen and chest, restricting breathing. The skin can crack, opening her up to infections.

The Foundation for Ichthyosis and Related Skin Types Inc., knows of two other children in the United States with the same disorder, a girl and a boy in San Diego and Houston.

No child with Harlequin ichthyosis has lived beyond her teens, most die within a few months of birth.



Hunter gets more than 15 topical medications to keep her skin from becoming infected or too dry.

Yet because of the concentrated efforts by her parents and professionals, the cheerful, active 18-month-old has defied all medical and social presumptions.

She loves the book "The Little Engine That Could." She can crawl and walk as unsteadily as any toddler.

Two weeks ago, she amused a visitor with her royal Queen's wave. The gentle twist of a raised right wrist that comes so easily to everyone else. Hunter had to be taught. The skin is so tight around her wrist that mobility can be painful.

"Hi! Hi!" she shouts into a telephone receiver with gusto. Then she laughs, straight from the belly.

Her tremendous progress is a result of concentrated health care and diligent professional help in movement and speech — which has cost more than \$300,000 in federal, state and private insurance funds during Hunter's young life.

More than half of the total, which accounts for Hunter's nursing care, is paid mostly by federal and state Medicaid funding.

icaid funding.

Patti's private health insurance with an HMO pays for Hunter's visits to six specialists, hospitalization, medical procedures and some medications.

That doesn't include the electric and water bills, which have tripled since she was born, and the \$100 a month for over-the-counter topical ointments.

And now, as federal and state governments race to cut their outlays for medical services, the consequences for children like Hunter are becoming all too apparent.

First, the Republican-controlled Congress wants to restrict the definition of disability. If the categories of eligible medical assistance are restricted, Hunter could lose her disability classification.

She also could lose her medical assistance because Pennsylvania may use parental income as part of the criteria. While specific salary levels have not been determined, advocates fear that the Steintzes' income of just under \$47,000 last year could be too high for eligibility.

Then there is the early intervention program for which children like Hunter receive special services, such as physical, speech and occupational therapies.

Pennsylvania receives \$12.5 million a year from the federal government to support these services. But there's a catch: No child can be turned down.

Gov. Ridge, however, wants the state to determine whether a child is eligible. He also wants to cap spending for the children who receive the services. That's because it costs the state \$42.5 million more a year than what it receives from the feds to provide the services, or about \$8,000 a year for each of the estimated 16,000 Pennsylvania children who receive the services, according to the state Welfare Department.

SEE HUNTER, PAGE F.14



Patti shares a tender moment with Hunter during one of the child's two daily baths. Bath time can be a relief to Hunter because the warm water makes her skin smooth and less itchy.

Hunter's battle

HUNTER FROM PAGE F-13

"It's going to double in a five-year period, and it has a lot of people concerned, which is why the governor wants out — because we can't control the cost per child," Nancy Thaler, deputy secretary of the state Department of Public Welfare's Office of Mental Retardation, said at a hearing in Oakland.

It's not that parents like Patti and Mark view government help as a bottomless pit.

They see it as a means of helping families help their children become productive adults.

"Which way does the government want it?" asked Patti.

"We'll have to quit our jobs and go on welfare. I don't see any other way out." That is not an option they want to explore.

At stake, too, could be other federal funding that enables Hunter to receive the nutritional supplements her 17-pound body craves. She has digestive problems not directly related to the Harlequin disorder, but ones that interfere with her getting enough essential nutrition.

In recent days she has worried doctors because she has been unable to keep her food down.

"Nobody can understand what we go through," Patti recently told state Rep. Don Walko, D-North Side. "I fight for her every day. I fight with the insurance company. I'm always fighting with someone."

It took seven weeks for Hunter to make her first trip home. After three weeks at Magee, including the first nervous meetings between Hunter and her older sister, Dana, she spent another four weeks in a transitional unit in Shadyside.

Every midnight a nurse arrives at their 2½-story home to administer medicated eyedrops, take Hunter's temperature and pulse, and rub her body with at least one of the 15 topical ointments — all every two hours. The humidifier must be monitored also.

"We deal with 12 hours a day spent on Hunter's care alone," Patti said. "We juggle two full-time work schedules, Hunter's routine and doctor appointments."

"Thank God I have a wonderful boss who allows me flexibility in my work hours to care for Hunter. Even so, my husband and I take days off work for doctor appointments, and I take days off to spend time with my forgotten child, Dana, who loves her sister so much."

Dana is also acting out. Her grades have dropped in school and is angry with me over things I cannot change. I cannot fix Hunter so that we can live a normal life.

"I don't know if parents of normal children can appreciate as much as we do how truly thrilling Hunter's accomplishments are."

Her voice choking, she added: "She played with her first friend last week."

More than anything in the world, Patti and Mark strive for normalcy in the frenzy around them.

For now, Mark is home Sundays and Mondays. Patti is off Thursdays and Fridays. On the other days, nurses take over.

Once a week, medical personnel come to the house to evaluate Hunter's progress. Other days, there are a variety of therapists working with her coordination, dexterity and other physical motions.

Every day, Hunter must be bathed in the morning and evening — each bath an up to two-hour event so her skin can be carefully smoothed and the extra skin peeled from inside her nose and ears.

"The evening bath has become a joint effort. It's something we do together. It's normal, though for me normalcy has changed a lot," Mark said.

While Mark fills the tub and spreads oversized bath towels on the floor in Hunter's bedroom, Patti makes a game with Hunter to climb the stairs to the second floor. She switches on lively music and spreads a plethora of lotions, ointments and other medications around her.

Hunter bounces to the music. She loves the warm water, the only part of the day when her skin doesn't itch. She squeals at the water pic, grabbing at the sensation of the waterfall that squirts into her ears. She drinks from it and gurgles.

Often, Dana joins in, splashing water on her baby sister.

Mark holds her; Patti gently scrapes away the peeling skin. She uses a special instrument for her ears and nose. Hunter becomes pinker and pinker as her new skin glistens in the bath water.

Afterward, looking shiny from the ointment and petroleum jelly covering her, Hunter proceeds with her nighttime ritual of saying good-night to her lamp and her clock and her stuffed animals.

While Mark empties the bathwater, Patti gently rocks Hunter and puts her into the crib. It's 10 p.m.

In two hours, the overnight nurse will arrive and her parents can go to bed.

And then begin their day again, tomorrow.

... when it can

By Ann Rodgers-Melnick
Post-Gazette Staff Writer

"Uh, yeah. Father, me and my buddies — we all was altar boys down at St. Joe's in the old days — and we got this bet. Is it OK to eat turtle soup on Fridays in Lent?"

The Rev. Lawrence DiNardo has heard that one before. His office for canonical services in the Catholic Diocese of Pittsburgh fields 25 to 30 calls a day from people with questions about church law, policy and doctrine. Some are urgent and se-

the Mass was in Latin. Bosco says callers have become more conservative over the years, often protesting church reforms. DiNardo mostly notices that they have become more argumentative.

"Every once in a while you get a call that blows your mind. You feel that you are talking to a person who needs help, but you don't know what to do," DiNardo said.

One caller claims her husband is imprisoned in a vat of whipped cream in the basement of St. Mary of Mercy, Downtown. He will be freed only if Bishop Donald W. Wuerl issues a decree.

SEE **CALLERS**, PAGE A-22

cu a ready has states scrimping

By Peter T. Kilborn
The New York Times

COLUMBIA, Mo. — A little-noticed provision in the nation's new welfare law that will immediately cut federal money for safety-net services like soup kitchens and child-protection programs has set off a scramble among state officials to preserve what they can.

As of Oct. 1, the government will chop 15 percent, or \$420 million, from the \$2.8 billion it distributes annually to the states in no-strings-attached money under a program known as the Social Services Block Grant.

States use the appropriations for hundreds of purposes — typically, to help the elderly poor stay out of nursing homes, pay for shelters and day care, rehabilitate juvenile criminals and rescue children from parental abuse or neglect.

States say the cuts come at the worst possible moment, when they expect the welfare law to start increasing demand for the services by people like legal immigrants, mentally disabled children and single adults who stand to lose food stamps or other assistance under the law's other provisions.

"It's not welfare reform," said Greg Vadner, director of the Division of Aging of Missouri's Department of Social Services. "You just have less money. It's purely a funding cut."

Like many states, Missouri is asking managers of local safety-net programs to prepare to accommodate the reduction. Steve Renne, deputy director of the Department of Social Services in Jefferson City, said the Legislature would consider a special appropriation to offset the cuts, using savings from the decrease in people or welfare because of the healthy economy.

But some services will be cut, too. "Within the next 30 days, we will have to decide," Renne said. "Our legislative folk don't want to be in the position of picking up the loss when the federal government cuts a program."

In Vadner's division, the cuts mean new applicants for home-delivered meals will be turned away. "We will reduce the number of people we serve," he said.

The concern intensifies at the community level, where social workers, teachers, counselors and local charities come face to face with the poor.

The Salvation Army in Columbia, which receives only 2 percent of its money from public agencies and none under the block grant, is already reeling from the impact of

SEE **WELFARE**, PAGE A-2

Turnpike's high-tech network short-circuited

By Peter J. Shelly

Post-Gazette Harrisburg Correspondent

HARRISBURG — The Pennsylvania Turnpike Commission set out to take a ride on the information superhighway.

It detoured on a road to nowhere. Even that wasn't cheap. Records obtained last week show that the project has cost \$587,665 in public money to start, then abruptly stop.

The plan to make millions off a high-tech venture has ended up dead — for now — and maybe on the way to court. Here are the essentials of what happened:

The commission, intrigued by the money-making potential of high-tech information systems, decided to install a fiber-optic network along the turnpike. It would span 506 miles, and be leased to telephone, cable television and Internet businesses.

Big money would pour in, everyone said, and the commission certainly could use it. Since 1992, when tolls were last increased, turnpike operating expenses have grown almost twice as fast as revenues, a management study showed.

"Millions," one staff member said of the revenue the network was supposed to produce. "Hundreds of millions."

Dozens of companies expressed an interest in the project. In October 1994, the Armstrong Fiber Optic Network was selected after telling the commission it would install the network for free.

Armstrong would build the system at a cost of at least \$50 million. Then it would give the turnpike 16 lines for its own use. On top of that, the company was willing to give the commission a share of the profits made by leasing access to the network.

It didn't happen. Negotiations sputtered, and not one shovel of dirt was turned to get the project going.

Last week, the commissioners voted 3-2 to end negotiations with Armstrong.

Republican commissioners James Malone III, Robert Gleason Jr. and Bradley L. Mallory voted to start the superhighway project all over again.

Gleason did not vote on the project in 1994 because his insurance agency does business with Armstrong, he said last week. This time around, he cast the third vote to scuttle the project.

The only reason the Republicans offered publicly for their decision was that they wanted to give Robert Brown, hired as deputy executive director for information

SEE **PIKE**, PAGE A-22

Inside the Region

□ Patti Steinitz, center, and Mary Ann Veremeychic, a nurse with Interim Nursing, play peekaboo with Steinitz's daughter, Hunter.

Hunter is doing remarkably well for a 23-month-old with a genetic disease so rare that victims seldom live past 2 years. But now, her parents say Hunter's health could be threatened by cuts in the federal Supplemental Security Income program, which was included in the new welfare law that President Clinton signed in August.



Annie O'Neill/Post-Gazette

Story, Page B-5.

THE REGION & STATE

PITTSBURGH POST-GAZETTE ■ SUNDAY, SEPTEMBER 22, 1996

B-5



BRIAN O'NEILL

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Welfare cuts could be fatal to infant

By Ellen M. Perlmutter
Post-Gazette Staff Writer

Hunter is doing remarkably well for a 23-month-old with a genetic disease so rare that victims seldom live past 2 years.

But now, her parents say, Hunter's health could be threatened by cuts in the federal Supplemental Security Income program, which was included in the new welfare law that President Clinton signed in August.

Because of her SSI eligibility, Hunter Steinitz, who lives with her family in Ob-servatory Hill, has received almost \$200,000 in nursing care in her lifetime — one of the keys to her survival, say her parents, Patti and Mark Steinitz.

She is not the only one to get such ex-



Annie O'Neill/Post-Gazette

Hunter Steinitz — Within the next few weeks, she will have an operation to insert a tube in her belly so her parents can intravenously feed her the nutrients she needs.

traordinary help.

There are 115,000 children nationwide, including 15,000 in Pennsylvania, with severe physical and mental disabilities whose care may be in jeopardy under the revamped welfare law, advocates say.

Hunter, who was profiled in the Pitts-burgh Post-Gazette in May, has skin that grows so fast it can't shed quickly enough. Her skin becomes so thick that it tightens around her chest and abdomen, and could choke her. It also cracks easily, opening

her up to infections.

She needs some type of preventive medical care nearly every hour, whether it's moisturizing her skin or placing prescrip-tion drops in her eyes because her lids do not shut completely. A humidifier needs to be constantly monitored so the air is kept properly moist for Hunter's delicate skin — always bright pink as it grows and sheds.

Hunter requires two baths a day, each one taking up to two hours. Special care

must be taken to remove excess skin from her nostrils and ears.

The nurses, made possible through SSI eligibility, help Hunter's family to live an almost normal life. The couple, who also have a teen-age daughter, share the re-sponsibilities on their days off from work.

But, they said, they can't be available 24 hours a day.

"We need the nurses so desperately," Patti Steinitz said. "We have a night nurse every night of the week so we can get some sleep."

The nurses, paid through the state, are provided because of the severity of Hunt-er's disorder. An evaluation process en-abled Hunter to qualify.

But the new welfare law eliminates the assessment process Hunter went through. That process broadly defined a child's eli-gibility so that no children would fall through federal cracks.

Critics charged that the rule made it too easy for children to be classified with phys-ical or mental disabilities. They said par-ents coached their children to feign disabilities.

The General Accounting Office and So-cial Security Administration, however, de-termined there was "absolutely zero evidence of widespread abuse."

The new law requires SSA to come up with a new assessment procedure no later than Nov. 22.

SEE HUNTER, PAGE B-6

Standardized tests rapped

State assessment

SATURDAY NIGHT'S LOTTERY DRAWINGS HAD NOT BEEN CONDUCTED BY PRESS TIME.

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demically and how effectively schools are educating them.

It includes reading and math tests that are administered to fifth-, eighth- and 11th-graders, and writing exams that are given to sixth- and ninth-graders.

But the state test has been beleaguered with complaints from its inception.

"This test was designed to measure how well kids, and by extension schools, are doing at particular grade levels. We believe it doesn't do that," said Sean Duffy, press secretary for the state Education Department. "The material on the test is relatively rudimentary. We're going to move to assessment that is more rigorous."

even continue to take the current test.

Duffy said it was possible that an interim replacement exam would be given to youngsters next spring with a radically different one administered in 1998.

"We're just getting the data in for this test," Duffy said. "We haven't had a meeting on it yet."

Welfare cuts could be fatal

HUNTER FROM PAGE B-5

"Children with medically documented disabilities have a better chance of remaining eligible for benefits," said Tom Morgenau, SSA spokesman. That would seem to include Hunter.

But, said Jonathan Stein, general counsel of Community Legal Services in Philadelphia, "No one can give anybody any assurance."

"The fact is there is no standard right now. Since the law went into effect on Aug. 22, there has been no standard," Stein said. "We're in limbo."

Meanwhile, Hunter's parents juggle their schedules to deal with Hunter's condition. Her mother is a Pittsburgh police officer who made about \$40,000 last year. Her father is a struggling businessman who recently closed his framing shop on the South Side.

Their income was not taken into consideration in determining SSI eligibility — only their daughter's extreme need for care.

Hunter weighs 17 pounds, 3 ounces now — 12 pounds more than her birth weight. She has a digestive problem that makes it difficult to keep food down.

Within the next few weeks, Hunter will have an operation to insert a tube in her belly so her parents can intravenously feed her the nutrients she sorely needs.

Patti Steinitz's private medical insurance pays for some medicines and care. But it has caps on the amount it will disperse in Hunter's

lifetime.

She was born with the disorder as a result of two rare, recessive genes carried by her parents. The condition is present in one to five children of every 5 million babies born in the United States each year. There are only three children, including Hunter, in the United States diagnosed with Harlequin Ichthyosis.

"Without [SSI], her care would decline and leave her at risk to illnesses and death," Patti Steinitz wrote to Clinton a month before he signed the welfare bill.

The SSI changes can also affect Hunter's eligibility for other programs that help with her medical care.

They include the federal Women, Infants and Children program, which provides the nutritional formulas she needs to ensure she gets the 1,300 calories a day her body craves. And because many state programs are predicated on SSI eligibility, the new law could also affect whether the state would continue to pay for early intervention services — such as physical therapy — that Hunter now receives.

The toddler, who could barely lift her head her first year, is as active and inquisitive as any healthy child her age because of the constant care she receives, her parents maintain.

"I wish people would understand that this is not something we chose to happen to us," Patti Steinitz said. "It happened. We love our baby. And we're going to do everything we can for her. Everything."

PG series on mentally ill wins award

A two-part series titled "Prisoners of the Mind" by Pittsburgh Post-Gazette writer Barbara White Stack and photographer Martha Rial has received the 1996 media award from the Alliance for the Mentally Ill of Pennsylvania.

The series, which ran in November 1995, focused on a mentally ill woman who was institutionalized for 50 years before being released to her daughter.

White Stack and Rial spent 15 months with Bernice Hensel and her mother, Marian Deibaugh, after Deibaugh's release from Mayview State Hospital, tracking the tribulations Hensel encountered in caring for her mother.

Thomas V. Gargan, president of the AMI, said their work "inspired others and has led both mental health professionals and the general public toward the truth about serious mental illness and away from ignorance and the stigma associated with brain disorders."

The award will be presented at the AMI conference in Grantville, Dauphin County, Oct. 5.

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Informational Childhood Disability Notices

SSA will be sending notices to approximately 340,000 disabled children (and their payees) who are now receiving Supplemental Security Income benefits. The purpose of the notices is to explain how recent legislation may affect those benefits.

- Informational Notice to Disabled Children Who Are or Will Be Age 18 by 9/30/97

About 60,000 of these children become age 18 during the period 8/22/96 - 9/30/97. If they are still receiving SSI at age 18, we must redetermine their eligibility under adult rules for disability. Since some of these recipients are already age 18, we need to review their cases as soon as possible. For this reason, we plan to send the Age 18 Childhood Disability Notice to each of these 62,000 children by early December 1996. This is the notice that is now being submitted to the Domestic Policy Council for review. It explains why the children's cases must be redetermined and what rules will be used for the redeterminations.

- Informational Notice to Other Disabled Children

The remaining 280,000 children will have their eligibility redetermined under the new childhood disability rules. These children will receive the other informational notice, which was reviewed by the Domestic Policy Council last week. That notice is also expected to be mailed by early December 1996.

No. 50,625

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U.S. to Review Disability Aid For Children

Welfare Law Imperils Benefits for 260,000

By ROBERT PEAR

WASHINGTON, Nov. 27 — The Government sent letters to the parents of 260,000 children today notifying them that the children might lose disability benefits because of the new welfare law.

The cash benefits, averaging \$424 a month, are paid under the Supplemental Security Income program. The children, most from low-income families, were previously found to have a wide variety of severe physical or mental disabilities like cerebral palsy, autism, tuberculosis, diabetes, sickle cell anemia, epilepsy and mental retardation.

Some children, especially those with mental, emotional and behavioral problems, will lose the cash benefits they now receive. Some will also lose Medicaid coverage.

For years, critics of the fast-growing program have contended that it was too generous and that some children received benefits even though their medical problems were not very severe. The program was a prime target of Republicans who wrote the welfare bill, which President Clinton signed on Aug. 22.

Philip A. Gambino, a spokesman for the Social Security Administration, said the letters were being sent to 260,000 of the one million children who receive disability benefits. The Government will review the cases to see if the youngsters meet the definition of disability established by the new law and regulations.

Mr. Gambino estimated that 100,000 to 200,000 children — 10 percent to 20 percent of all children on the rolls — would eventually lose benefits.

The effort begun today by the Government will be a test of resolve for those determined to prune the rolls and for others trying to minimize the

U.S. Reviews Disability Aid Imperiled by Welfare Law

Continued From Page A1

impact of the new welfare law. Past attempts to reduce the number of children in the program encountered stiff opposition.

The new law tightens eligibility standards, but Congress gave the President great latitude to interpret the law. Administration officials are writing regulations that will, by redefining disability, do much to determine the impact of the law. The new regulations are supposed to be issued in the next few weeks. Administration officials said Mr. Clinton would probably review the rules because the decision could set off a political furor.

In a sample letter to parents, Janice L. Warden, a Deputy Commissioner of Social Security, said, "We will review Jane Doe's case to see if she is disabled under the new definition of disability for children." And if we find she is not disabled because of the new law, we will stop her S.I., unless you appeal our decision," it said.

The new law does not stop benefits immediately. Children now on the rolls may receive benefits through June 1997, provided they continue to meet all the other eligibility criteria.

Parents of disabled children and their advocates urged Mr. Clinton to be compassionate as he applies the law.

"The power to hurt these children, or to assist them, is completely in President Clinton's hands," said Martha E. Ford, a lobbyist for The Arc, formerly known as the Association for Retarded Citizens.

Children qualify for disability benefits only if their parents' income and assets are less than certain levels set by law. Benefits decline as family income rises. Families generally become ineligible as incomes approach \$30,000 a year. Two-thirds of the disabled children receiving benefits are in families with low incomes — less than \$21,000 a year for a family of four in 1995.

Under the new law, a person under the age of 18 is eligible for benefits if he or she has "a medically determin-

A CLOSER LOOK

The Benefits

WHAT One million children receive disability benefits under the Supplemental Security Income program, created by Congress in 1972. The average monthly benefit is now \$424.

WHO About two-thirds of children on the rolls are eligible for benefits because of mental impairments. Nearly two-thirds of these youngsters, 42 percent of the total, are mentally retarded.

The most common physical disabilities include leukemia, asthma, Down syndrome, deafness, blindness, epilepsy and cerebral palsy. Recipients must meet certain income eligibility standards.

WHY The number of children on the rolls has tripled since 1989, and costs have quadrupled, to slightly more than \$5 billion a year.

Enrollment grew because of court decisions, changes in eligibility criteria, an increase in the childhood poverty rate and a Congressional mandate to locate children who might qualify for benefits.

Sources: Social Security Administration, National Commission on Childhood Disability.

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"Kennedy is an all-or-nothing airport," Mr. Monaghan said of the traffic delays on the ground.

Controllers do have some now

able physical or mental impairment, which results in marked and severe functional limitations." The law does not define "severe," but in a report on the bill, House and Senate negotiators said the term was to be given "its common-sense meaning."

Under the old law, children could qualify for disability benefits in two ways: if they met the medical criteria in a list of impairments, or if they were unable to engage in activities appropriate for their ages. The new law eliminates the second alternative, which used an "individualized functional assessment" for each applicant.

tools to handle all the traffic. One is a surface radar, perched directly atop the tower in a giant golf ball, which shows the outline of objects on the axiways and runways. The initial impression of objects on the screen is like bugs under a large rock: some large, with plane-like outlines, representing big jets, and some tiny, probably panel trucks or other small vehicles, all moving at different speeds. And there is the tower itself, two old and 100 feet taller than the one. Through its tinted windows, controllers can see everything in a 360-degree arc. Recently, they spotted one in the parking lot breaking a controller's new car.

J. Johnson, a supervisor of the thief was foiled by a controller, who diverted a plane to hover over the car until it arrived.

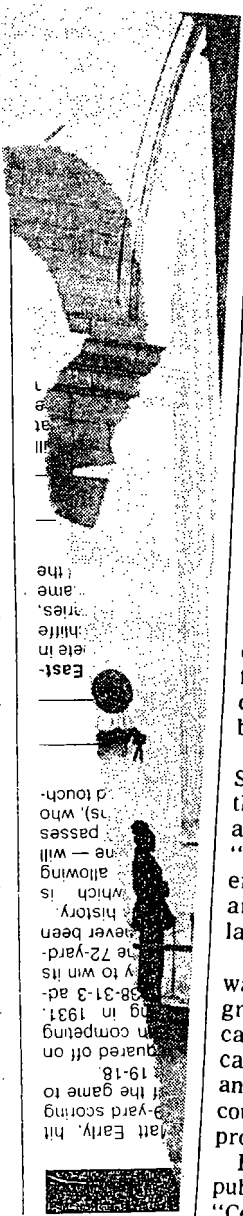
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The children receiving notices this week got on the rolls because of such assessments. The new law says that the Government must review the eligibility of all children on the rolls at least once every three years, unless a child's condition is considered unlikely to improve.

The language of the new law shows that Congress still wanted to pay benefits to children with "severe functional limitations," suggesting that the Government still must assess the effects of disabilities and chronic illnesses on children's lives. But Congress did not say how severe a disability must be.

The Senate Democratic leader, Tom Daschle of South Dakota, said, "Congress provided the executive branch with great latitude to interpret the statute."

Discussing the legislation when he was a Senator, Bob Dole said the Government must "obtain a realistic picture of how an impairment affects each child's abilities."

Comments by Mr. Dole and other lawmakers show that they wanted the Social Security Administration to use some kind of functional test that would go beyond a simple listing of the more common impairments. Such tests are needed to evaluate children who have multiple impairments or rare disorders that limit their ability to perform everyday activities, Mr. Dole said.

The number of children receiving disability benefits has increased sharply, to 1 million today from 290,000 in 1988. Much of the increase has occurred since 1990, when the Supreme Court ruled that the Government had violated Federal law by using overly strict standards.

In the fiscal year that ended Sept. 30, the Federal Government spent \$26 billion on the Supplemental Security Income program, including a little more than \$5 billion for children. The Congressional Budget Office says the new restrictions on children's benefits will save a total of \$7 billion in the next six years.

But Mr. Gambino said that Social Security officials were not writing the new rules to save any specific amount of money and had given "very little" weight to fiscal considerations. The agency's goals, he said, are to carry out the spirit of the new law and to protect children.

Just how drastically Congress wanted to change the disability program is in dispute. House Republicans originally wanted to abolish cash payments for disabled children and replace them with vouchers that could be spent on a more limited program of medical care.

But Senator William S. Cohen, Republican of Maine, said recently that "Congress is not demanding a drastic change in the level of severity required" to qualify for benefits.

Mr. Dole, whose right arm was ruined by war injuries, said last year the program "can be a lifesaver" for families with disabled children.

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November 15, 1996

To: Those Concerned with the Adoption of a New
SSI Child Disability Policy

The attached new standard drafted to comply with and implement the new welfare law's SSI child disability provisions, has as its thrust insuring that SSI is provided only to truly disabled children through a revised test that emphasizes accuracy, reliability, and thoroughness in adjudication of claims. This proposal adds to and refines a proposal submitted earlier by a coalition of child disability advocates.

Among its hallmarks are:

1. So-called "moderate" scores in functioning must now, as a result of proposed regulations, be "good and solid moderates" where the disability "interferes considerably" with the child's ability to function like other children that age.

A corollary rule will be that not all "three moderates" will require an allowance of the claim.

2. There will no longer be a binding rule that "one marked and one moderate" will dictate an allowance; and the "solid" moderate rule will also be applicable in these decisions.

3. New rules will avoid any double-counting in domains of functioning that might have given too much laxity in the former IFA.

4. Evidentiary development will be emphasized in many areas, especially to document in a reliable way the underlying medical diagnosis, to provide longitudinal evidence of the history of the child's impairments, and to insure consistency between standardized testing employed and evidence of functional limitations.

5. Fairer evaluations of physically disabled children disfavored by the old IFA are provided by an additional domain relevant to their disabling conditions.

Jonathan M. Stein
General Counsel
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November 15, 1996

NEW CHILDREN'S SSI DISABILITY STANDARD

By Thomas Yates

In the SSI Coalition Proposal For A New Children's SSI Disability Standard (Draft, September 4, 1996) ("SSIC Proposal"), we, and a coalition of national disability groups, laid out a proposal for determining children's SSI disability, that charted a middle course between the Individualized Functional Assessment test that was rejected by the welfare reform bill and sole reliance on the Listings step (by meeting or equalling its criteria) of the children's disability sequential evaluation.¹

Our proposal set forth a new childhood disability sequential evaluation that retained the current first three steps, see 20 C.F.R. §§ 416.924(c)-(e), and incorporated a new step four that specifically focused on whether a child had medically determinable impairments that caused marked and severe functional limitations.²

The new fourth step is three-pronged. The first prong makes clear that all relevant evidence, both medical and non-medical,

¹ Sole reliance on the Listings was also rejected by Congress. The House of Representatives passed a version of a welfare bill that made the present Listings step the childhood disability standard. The Senate rejected that approach, and substituted a childhood disability standard that found children disabled if their medical impairment(s) caused marked and severe functional limitations. The Senate version was then adopted by the House in the conference bill.

² The SSIC Proposal is significantly different from the discarded IFA test because it tightens the disability threshold test. The IFA provided that generally, but not invariably, children with three moderate impairments would be found disabled, and it disfavored children with physical disabilities. The SSIC Proposal does not so provide; additional restrictions added to that proposal in this document further distance the SSIC proposal from the IFA test.

must be considered in determining whether a child's impairments cause marked and severe functional limitations.

The second prong requires decisionmakers to determine whether children have deficits in development or functioning in up to seven different areas. Those areas, which are modelled on the "B" criteria of the present childhood mental impairment listings, are: cognitive functioning; communicative functioning; social functioning; personal/behavioral functioning; gross and fine motor functioning; physical stamina and basic physical functioning; concentration, persistence, and pace; and responsiveness to stimuli (for infants).

The third, and eligibility determining, prong provided that a child would be found to have "marked and severe functional limitations" if his or her limitations in the areas listed above showed: a) inability to perform substantially any function; b) a marked impairment in one area and a moderate impairment in another; or c) a combination of impairments in the various areas that caused marked and severe functional limitations.

In this document, we supplement that proposal, and in doing so further tighten the disability standard, by addressing the following three issues that are intended to respond to Congressional concerns:³

- clarifying the disability threshold set by the proposed three tests of disability listed above to ensure that only children with marked and severe functional limitations are found disabled;
- ensuring that decisionmakers do not double-count functional limitations as they assess a child's development or functioning in the areas listed above; and
- defining with greater specificity what evidentiary showing is required to show marked or moderate impairments in the relevant areas.

A. Clarifying The Disability Threshold Set By The Three Tests Of

³ We believe these Congressional concerns fall into three areas: a) the old IFA test was deficient because it allowed nearly all children with three moderate impairments to be found disabled (leading to disability findings for children who were not truly disabled); b) there was concern that some functional limitations caused by medical impairments were double-counted in assessing loss of functioning in various areas; and c) there were inadequate safeguards to ensure that adequate and reliable evidence was developed to ensure that children truly had certain impairments (i.e., the children were not malingering, cheating, being coached).

Disability For Marked And Severe Functional Limitations

We propose that a child would be found disabled with marked and severe functional limitations if any of the three following criteria were satisfied:

- impairments that prevent the child from performing substantially any function (e.g. walking, breathing, taking care of personal needs) contained in any one of the areas;
- a marked impairment in one of the areas and a moderate impairment in another; or
- a combination of impairments in the various areas that causes substantial reduction in the child's ability to function independently outside of the home within age-appropriate norms, or causes substantial reduction in the ability to function independently, appropriately, and effectively in an age-appropriate manner.

1. Background: Defining Degrees Of Functional Limitation

SSA assesses the severity of functional limitations caused by medical impairments on a scale of "extreme," "marked," "moderate," "mild," and "minimal." Understanding these gradations is essential to applying the three criteria set forth above, particularly because SSA defines "moderate" to be more severe than it is in common usage. The different gradations are:

SEVERITY OF FUNCTIONAL LIMITATION	DEGREE OF FUNCTIONAL LOSS
Extreme	"Extreme" means loss so severe that child is clearly disabled by the limitation(s); the limitation(s) meets or equals a listing (e.g. marked inability to stand or walk; frequent need for life-sustaining device at home or elsewhere; requirement for 24-hour-a-day supervision for medical or behavioral reasons)

Marked	"Marked" means more than "moderate" but less than "extreme." A marked limitation is one that interferes seriously with the child's ability to function independently, appropriately, and effectively in an age-appropriate manner and, when applicable, on a sustained basis. When standardized tests are used as the measure of a child's functional abilities, a valid score that is two standard deviations below the norm for the test (e.g., an IQ score of 70 or lower on the WISC-R) will be considered a marked restriction.
Moderate	"Moderate" means more than "mild" but less than "marked." A moderate impairment is one that interferes considerably with the child's ability to function independently, appropriately, and effectively in an age-appropriate manner, and when applicable, on a sustained basis. When standardized tests are used as a measure of a child's functional abilities, a valid score that is between approximately one-and-one-half and two standard deviations below the norm for the test will generally be considered a moderate restriction (e.g. an IQ score on the WISC-R ranging from 71 through 77). The spectrum of what may constitute "moderate" limitation in this age group ranges from limitations that may be close to the "marked" level in severity to limitations that may be close to the "mild" level and, thus, considerably less limiting.⁴
Mild	"Mild" means more than "minimal or no impact" but less than "moderate." A "mild" impairment is one that is "severe," as defined at 20 C.F.R. § 416.924(d); it causes more than minimal limitation in a child's ability to function independently, appropriately, and effectively in an age-appropriate manner.

⁴ In this point, the definition of "moderate" is more severe than SSA has represented that it is. In its Disability Digest No. 94-6 (March 1996), SSA stated that a "moderate" impairment could range from one that is almost "marked" to one that is almost "minimal." This proposal provides a higher low threshold for "moderate" impairments.

Minimal or No Impact	"Minimal or no impact" means that it would not be considered a "severe" impairment. The impairment or combination of impairments causes no more than minimal limitation in a child's ability to function independently, appropriately, and effectively in an age-appropriate manner.
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In making a disability determination, the decisionmaker must review a child's medically determinable impairments and assess the degree of functional limitations, using the definitions set forth above, in each of the relevant areas (e.g. cognition; communication; social functioning; personal/behavioral functioning; motor skills; concentration, persistence, and pace; physical stamina and basic physical functioning; and responsiveness to stimuli for infants).

2. Applying The Proposed Marked And Severe Disability Threshold Tests

In applying any of the three tests of marked and severe disability, the standards set forth are guidelines, not binding rules. Thus, children who meet their terms will generally, but not invariably, be found disabled. In determining childhood disability, SSA must act, on a case-by-case basis, to ensure that only children with marked and severe functional limitations are found disabled.

a. Inability To Perform Substantially Any Function Test

If a child has an extreme functional limitation in any of the areas, see examples set forth in table above, she or he would be considered disabled.

b. The One Marked And One Moderate Test

Generally, but not invariably, a child with one "marked" impairment and one "moderate" impairment will be found disabled. However, not all children found to have one "marked" impairment and one "moderate" impairment are necessarily disabled. Because the term "moderate" represents a range of limitations from just above "mild" to just below "marked," an impairment(s) that imposes "moderate" limitations in an area when paired with another "marked" impairment will not always lead to a finding of disability.

A "moderate" impairment, when paired with another "marked" impairment, cannot be considered disabling unless it is a "good, solid moderate." An impairment, that while in the "moderate" range, is just above "mild" will not be considered to be a "good solid moderate."

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Thus, the decisionmaker, in making a decision under this test, must assess the effects of the child's impairments on the various relevant areas, see § 4(b) of SSIC Proposal, and the "other factors" set forth at 20 C.F.R. § 416.924c, and before finding a "good, solid moderate" impairment:

- take a step back,
- look at the totality of the evidence, and;
- consider what the child is able to do and in what ways the child's impairment(s) interfere considerably with his or her age-appropriate functioning.

If the impact of the functional limitation interferes considerably with the child's functioning, then it is a "good, solid moderate" impairment that could support, when paired with another "marked" impairment, a finding of disability.

c. Combination Of Impairments In Various Areas Test

Finally, a child could be found disabled at the "marked and severe" functional limitations step if she or he has:

a combination of impairments in the various areas that causes substantial reduction in the child's ability to function independently outside of the home within age-appropriate norms, or causes substantial reduction in the ability to function independently, appropriately, and effectively in an age-appropriate manner.

This test, unlike the first two tests, sets forth no general standard in regard to the number of required marked or moderate functional limitations. It is intended to address those cases of severe disability in which a child evidences impairments in a number of the relevant areas that, taken in totality, cause "marked and severe" functional limitations.

Examples of cases in which children may be found disabled under this test are children with "good, solid moderate limitations" in at least three or more of the relevant areas, determined on a case-by-case basis. *Given that this revised definition*

This test is narrowly drawn to provide SSA decisionmakers with flexibility to make favorable disability determinations for children with functional limitations of equivalent impact to children found disabled with one marked impairment and one good, solid moderate impairment.

6 of "moderate" is more restrictive than that previously used, children with three moderate impairments in areas of functioning will no longer be automatically determined eligible.

B. Decisionmakers Must Ensure That Functional Limitations Are Not Double-Counted In Assessing A Child's Development Or Functioning In The Various Areas

1. Statement of Policy

The areas for assessment of functional limitations⁶ are intended to aid decisionmakers by dividing various areas of a child's functioning or development into different and unique groupings to ensure that functional limitations are not double counted in determining childhood disability.

Thus, decisionmakers must ensure that any functional limitation that a child has is not assessed in more than one area in determining disability. For example, a child's reading difficulties are evidence of a cognitive limitation, but not a communication limitation--no impairment should be found in communication as a result. In addition, a child's inability to relate satisfactorily to others and his or her difficulty and frustration in not being able to relate satisfactorily to others constitutes a limitation in the social area, not the personal/behavioral area.

Set forth below are definitions of the various areas. Decisionmakers must ensure that any functional limitation caused by medically determinable impairments are assessed in only one area.⁶

² The relevant areas are as follows:

for a child from birth until attainment of age 1: cognitive functioning; communicative functioning; ability to sustain social interaction on an ongoing reciprocal fashion; motor development; age-appropriate physical stamina and basic physical functioning; and responsiveness to stimuli;

for a child from age 1 to attainment of age 3: cognitive functioning; communicative functioning; social functioning; gross and fine motor development; and physical stamina and basic physical functioning; and.

for a child from age 3 until the attainment of age 18: cognitive functioning; communicative functioning; social functioning; personal/behavioral functioning; gross and fine motor functioning; physical stamina and basic physical functioning; and age-appropriate concentration, persistence, or pace.

⁶ This does not mean that a medically determinable impairment (and resulting treatment) cannot cause more than one type of functional limitation. For example, a child with asthma could be

2. Definitions Of Various Areas:

a. Cognition

Cognition refers to the ability to learn through perception, reasoning, or intuition, and to retain, use, and manifest acquired knowledge in action or communication.

b. Communication

Communication refers:

- with respect to language, to the ability to receive, comprehend, and express messages using age-appropriate communication skills in order to meet one's needs, or to obtain or convey information; and
- with respect to speech, to audibility, intelligibility, and efficiency of speech production.

c. Fine And Gross Motor Skills Development

Fine and Gross Motor Skills Development refers to the ability to use one's body and extremities in gross and fine motions in order to relate to the physical environment and serve one's own or others' physical needs or purposes.

d. Personal/Behavioral Patterns

Personal/Behavioral ^{functioning} means age-appropriate activities and behaviors entailed in:

- self-help (e.g. feeding, dressing, maintaining personal hygiene);
- self-regulation (e.g. maintaining proper nutrition, sleep, health habits, regulating mood);
- self-improvement (e.g. increasing self-help behavior through learning of new skills);
- self-protection (e.g. taking necessary safety

limited in physical stamina as a result of the asthma and limited in personal/behavior functioning because of hyperactivity caused by medications used to treat the child's asthma. Similarly, a child with a history of otitis media with speech and language problems could have functional limitations in both communication (due to the hearing problems) and social (due to his inability to form, develop, and sustain relationships as a result of frustration resulting from the hearing and speech problems).

~~_____~~ In some cases, this means developing and considering evidence concerning the child's functioning that ~~_____~~ prior to the date of SSI application.

- SSA must when standardized tests are used to assess functional limitations, develop evidence of the child's day-to-day functioning in the relevant areas addressed by the standardized testing to ensure that test results are valid and consistent with the child's functioning.

covers a significant period of time.

- SSA must, when evaluating behaviors caused by medical impairments that impact functioning, obtain evidence regarding:

- the nature and intensity of the behavior--more is

While standardized intelligence tests are useful in assessing a child's cognitive abilities, they should not alone establish either marked or moderate limitations in cognitive functioning. While low scores on such tests are often indicative of significant cognitive limitations, they must be accompanied by achievement test scores when academic performance is an issue, and by reliable descriptive information about a child's cognitively based functioning on a daily basis. A child may function either above or below the level one would expect based on intelligence test scores alone.

Generally, impairment in cognitive functioning cannot be found unless a child manifests both subaverage general intellectual functioning and deficits in performing cognitively based tasks in an age-appropriate manner on a daily basis.

For example, a child with low IQ scores who nonetheless demonstrates relatively strong cognitive abilities in his or her day-to-day functioning may be found less limited than one would expect based on test scores alone. It is also possible for a child with higher IQ scores to be unable to perform cognitively based tasks in an age-appropriate manner on a daily basis. This child would be found more limited than one would expect based on test scores alone.

In assessing marked and moderate cognitive limitations, a child whose scores on standardized intelligence tests fall two standard deviations below the norm has a marked limitation, assuming the other relevant evidence shows corresponding limitations in day-to-day cognitive functioning. A child whose scores are between approximately one-and-one-half and two standard deviations below the norm for the test has a moderate restriction (e.g. an IQ score on the WISC-R ranging from 71 through 77), assuming consistency with other evidence.

required than a general allegation of the inappropriate behavior--by documenting a detailed description of what occurs, the precipitating event(s), and the setting(s) in which it occurs;

- the frequency and duration of the behaviors so that they can be assessed in longitudinal context and compared to the behaviors exhibited by "normal" children; and
- any interventions attempted to change the child's behaviors and the responses to these interventions.

In assessing any functional limitations resulting from such behavior, the decisionmaker should decide whether the behaviors are consistent with any established impairment, whether there may be an additional underlying mental impairment which must be documented, and how and to what degree an established impairment affects the child's ability to function in an age-appropriate manner.

D. Conclusion

This proposal provides the best approach for SSA to implement the changes mandated by the welfare reform bill, and address the concerns raised by Congress. Its application will allow SSA to make childhood disability determinations that ensure that children with marked and severe disabilities are not improperly denied disability.