Family Practice Grand Rounds

Care of a Troubled Family and Their Child with Sickle Cell Anemia

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Cleveland, Ohio

In this Grand Rounds, a weekly educational activity in the Department of Family Medicine, Case Western Reserve University, the interrelationship of illness and family function is explored. This focus is firmly rooted in the patient care and educational perspective of this department. Today's presentation is unique only in that the presenters are second-year medical students.

MS. CAROL LEVITT (Second-year medical student): In presenting today's Grand Rounds, we would first like to begin with a brief background on Mr. and Mrs. B.

MR. MICHAEL FINE (Second-year medical student): Mr. and Mrs. B. are an attractive, young black couple who live in the inner city of a large, industrialized urban area. Both are high school graduates, and Mr. B. has had one year of college. Each carries the sickle cell trait, although sickle cell disease is more pronounced in Mr. B.'s family (Figure 1). Both Mrs. and Mrs. B.'s parents were divorced early in their childhoods, with their mothers remarrying. Each had difficulty relating to their families of origin and share a personal sense of inferiority.

The couple met in 1978 and had a stormy court-

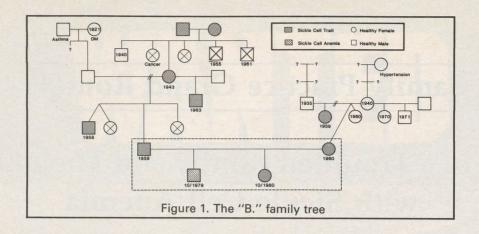
ship. They married with no family members in attendance when Mrs. B. was over five months pregnant. During their courtship, Mr. B. joined a conservative, fundamentalist church. Mrs. B. supports her husband's church involvement but is a more nominal member.

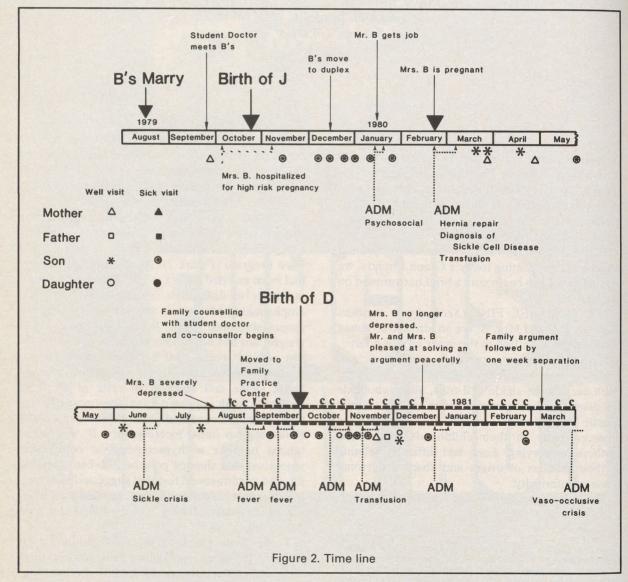
MS. LEVITT: I first met the B. family during Mrs. B.'s seventh month of pregnancy, when I was a first-year medical student in our Family Care Program (Figure 2). At that time the couple had been married for six weeks and were facing a multitude of difficulties, including Mr. B.'s unemployment. Moreover, the pregnancy was not progressing well. Mrs. B. had not gained any weight, nor had her fundus height increased for the last several weeks, and her blood pressure was gradually increasing. Mr. B. was openly worried about sickle cell disease. In spite of all these problems, both appeared mutually supportive and appropriately concerned about their unborn child. Following our initial meeting, Mrs. B. was hospitalized because of hypertension, a contraction stress test that showed poor beat-to-beat variability, and an ultrasound test that suggested intrauterine growth retardation. Labor was induced at 37 weeks' gestation. Jimmy, a 4 lb, 1 oz baby, was delivered.

From birth Jimmy was a difficult child to mother. He cried constantly, had chronic diarrhea, and spit up after every feeding. Before long, Jimmy's fussiness began to affect the couple's re-

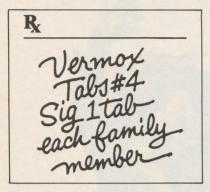
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DESCRIPTION VERMOX (mebendazole) is methyl 5-benzoylbenzimidazole-2-carbamate.

ACTIONS VERMOX exerts its anthelmintic effect by blocking glucose uptake by the susceptible helminths, thereby depleting the energy level until it becomes inadequate for survival. In man, approximately 2% of administered mebendazole is excreted in urine as unchanged drug or a primary metabolite. Following administration of 100 mg of mebendazole twice daily for three consecutive days, plasma levels of mebendazole and its primary metabolite, the 2-amine, never exceeded 0.03 $\mu \rm g/ml$ and 0.09 $\mu \rm g/ml$, respectively.

INDICATIONS VERMOX is indicated for the treatment of *Trichuris trichiura* (whipworm), *Enterobius vermicularis* (pinworm), *Ascaris lumbricoides* (common roundworm), *Ancylostoma duodenale* (common hookworm), *Necator americanus* (American hookworm) in single or mixed infections. Efficacy varies as a function of such factors as pre-existing diarrhea and gastrointestinal transit time, degree of infection and helminth strains. Efficacy rates derived from various studies are shown in the table below:

	Whipworm	Common Roundworm	Hookworm	Pinworm
cure rates mean (range)	68% (61-75%)	98% (91-100%)	96%	95% (90-100%)
egg reduction mean (range)	93% (70-99%)	99.7% (99.5%-100%)	99.9%	

CONTRAINDICATIONS VERMOX is contraindicated in pregnant women (see Pregnancy Precautions) and in persons who have shown hypersensitivity to the drug.

PRECAUTIONS PREGNANCY: VERMOX has shown embryotoxic and teratogenic activity in pregnant rats at single oral doses as low as 10 mg/kg. Since VERMOX may have a risk of producing fetal damage if administered during pregnancy, it is contraindicated in pregnant women.

PEDIATRIC USE: The drug has not been extensively studied in children under two years; therefore, in the treatment of children under two years the relative benefit/risk should be considered.

ADVERSE REACTIONS Transient symptoms of abdominal pain and diarrhea have occurred in cases of massive infection and expulsion of worms.

DOSAGE AND ADMINISTRATION The same dosage schedule applies to children and adults. The tablet may be chewed, swallowed or crushed and mixed with food. For the control of pinworm (enterobiasis), a single tablet is administered orally, one time. For the control of common roundworm (ascariasis), whipworm (trichuriasis), and hookworm infection, one tablet of VERMOX is administered, orally, morning and evening, on three consecutive days. If the patient is not cured three weeks after treatment, a second course of treatment is advised. No special procedures, such as fasting or purging, are required.

HOW SUPPLIED VERMOX is available as chewable tablets, each containing 100 mg of mebendazole, and is supplied in boxes of twelve tablets. VERMOX (mebendazole) is an original product of Janssen Pharmaceutica, Belgium.

US Patent 3,657,267 December 1979

Committed to research... because so much remains to be done.

Tableted by Janssen Pharmaceutica, Beerse, Belgium for



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lationship, and they began arguing over the child's care.

Over the next several months, Jimmy had several hospitalizations for a variety of reasons (Figure 2) and numerous sick visits to the pediatric clinic. A visiting nurse also began seeing the family weekly. During the hospitalization for repair of an inguinal hernia at four months of age, a positive diagnosis of sickle cell anemia was made. During this time, Mrs. B. learned that she was pregnant again. Neither Mr. nor Mrs. B. felt they were ready for another child, but Mr. B.'s strong religious orientation prevented them from considering an abortion. During this period, the relationship showed increased signs of strain. In the presence of all of their problems (sick and difficult child, an unwanted pregnancy, and a troubled marriage), when I asked her what her biggest worry was, Mrs. B. said she most wanted help with her marriage. I tried to arrange marital therapy for the couple but was unsuccessful. Mrs. B. became increasingly depressed, and in August, when she was seven months pregnant, I sought psychiatric intervention. This also failed. At this point I decided to try talking to Mr. and Mrs. B. together. They were responsive to my initial attempts, but before long I found myself siding with Mrs. B. during our meetings. With the B.'s approval, I invited Mr. Michael Fine, who is also a medical student and my husband, to join us in the marital sessions. I felt that with another man present, the sessions would be more balanced.

One month later, in September 1980, Mr. B. obtained health insurance because he was now working, and the family became members of the Family Practice Center under a prepaid health plan. At the Family Practice Center, a protocol to deal with Jimmy's physical condition and the B.'s psychological situation was developed. My involvement, as a medical student with the family, would continue now with supervision from the family practice faculty for both the B.'s emotional and medical care. Consultation regarding Jimmy's sickle cell disease would be provided by the pediatric hematology department.

September continued to be very stressful for the couple, with Jimmy being very irritable and having

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THE JOURNAL OF FAMILY PRACTICE, VOL. 15, NO. 1, 1982

Brief Summary: Before prescribing, please consult complete prescribing information, a summary of which follows:

Indications: IMODIUM is indicated for the control and symptomatic relief of acute nonspecific diarrhea and of chronic diarrhea associated with inflammatory bowel disease. IMODIUM is also indicated for reducing the volume of discharge from ileostomies.

Contraindications: IMODIUM is contraindicated in patients with known hypersensitivity to the drug and in those in whom constipation must be avoided.

Warnings: Antiperistaltic agents should not be used in acute diarrhea associated with organisms that penetrate the intestinal mucosa, e.g., enteroinvasive <u>E. coli, Salmonella, Shigella</u>, and in pseudomembranous colitis associated with broad-spectrum antibiotics.

Fluid and electrolyte depletion may occur in patients who have diarrhea. The use of IMODIUM does not preclude the administration of appropriate fluid and electrolyte therapy. In some patients with acute ulcerative colitis, agents which inhibit intestinal motility or delay intestinal transit time have been reported to induce toxic megacolon. IMODIUM therapy should be discontinued promptly if abdominal distention occurs or if other untoward symptoms develop in patients with acute ulcerative colitis.

Precautions: In acute diarrhea, if clinical improvement is not observed in 48 hours, the administration of IMODIUM should be discontinued. Abuse and Dependence: Physical dependence to IMODIUM in humans has not been observed. However, studies in monkeys demonstrated that loperamide hydrochloride at high doses produced symptoms of physical dependence of the morphine type.

Carcinogenesis: In an 18-month rat study with doses up to 133 times the maximum human dose (on a mg/kg basis) there was no evidence of carcinogenesis.

<u>Pregnancy</u>: Safe use of IMODIUM during pregnancy has not been established. Reproduction studies performed in rats and rabbits with dosage levels up to 30 times the human therapeutic dose did not demonstrate evidence of impaired fertility or harm to the offspring due to IMODIUM. Higher doses impaired maternal and neonate survival, but no dose level up to 30 times the human dose demonstrated teratogenicity. Such experience cannot exclude the possibility of damage to the fetus. IMODIUM should be used in pregnant women only when clearly needed.

Nursing Mothers: It is not known whether IMODIUM is excreted in human milk. As a general rule, nursing should not be undertaken while a patient is on a drug since many drugs are excreted in human milk.

<u>Pediatric Use</u>: Safety and effectiveness in children have not been established. Therefore, use of IMODIUM is not recommended in the pediatric age group (under the age of 12). In case of accidental ingestion of IMODIUM by children, see Overdosage Section for suggested treatment.

Adverse Reactions: The adverse effects reported during clinical investigations of IMODIUM are difficult to distinguish from symptoms associated with the diarrheal syndrome. Adverse experiences recorded during clinical studies with IMODIUM were generally of a minor and self-limiting nature. They were more commonly observed during the treatment of chronic diarrhea. The following patient complaints have been reported: Abdominal pain, distention or discomfort; constipation; drowsiness or dizziness; dry mouth; nausea and vomitins: tiredness.

Hypersensitivity reactions (including skin rash), however, have been reported with IMODIUM use.

Overdosage: Animal pharmacological and toxicological data indicate that overdosage in man may result in constipation, CNS depression, and gastrointestinal irritation. Clinical trials have demonstrated that a slurry of activated charcoal administered promptly after ingestion of loperamide hydrochloride can reduce the amount of drug which is absorbed into the systemic circulation by as much as ninefold. If vomiting occurs spontaneously upon ingestion, a slurry of 100 gms of activated charcoal should be administered orally as soon as fluids can be retained.

If vomiting has not occurred, gastric lavage should be performed followed by administration of 100 gms of the activated charcoal slurry through the gastric tube. In the event of overdosage, patients should be monitored for signs of CNS depression for at least 24 hours. If CNS depression is observed, naloxone may be administered. If responsive to naloxone, vital signs must be monitored carefully for recurrence of symptoms of drug overdose for at least 24 hours after the last dose of naloxone.

In view of the prolonged action of loperamide and the short duration (one to three hours) of naloxone, the patient must be monitored closely and treated repeatedly with naloxone as indicated. Based on the fact that relatively little drug is excreted in urine, forced diuresis is not expected to be effective for IMODIUM overdosage.

In clinical trials an adult who took three 20 mg doses within a 24-hour period was nauseated after the second dose and vomited after the third dose. In studies designed to examine the potential for side effects, intentional ingestion of up to 60 mg of loperamide hydrochloride in a single dose to healthy subjects resulted in no significant adverse effects.

How Supplied: IMODIUM is available as 2 mg capsules of loperamide hydrochloride. The capsules have a light green body and a dark green cap, with "ORTHO 1000" imprinted on one segment and "IMODIUM" on the other segment. IMODIUM capsules are supplied in bottles of 100 and 500.

IMODIUM (loperamide hydrochloride) is an original product of Janssen Pharmaceutica, Belgium, and co-developed by Ortho Pharmaceutical Corporation, Raritan, New Jersey.

U.S. Patent 3,714,159

FAMILY AND SICKLE CELL ANEMIA

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several hospital admissions for fever in which no sepsis was found. Mr. Fine and I continued to meet with them for counseling, which was now being closely supervised by the family medicine faculty.

In October Mrs. B. gave birth to Darla, a healthy, beautiful, 6 lb girl. Since that time, the family's overall condition has shown considerable improvement. Mrs. B.'s depression has lifted, and she has become more animated, communicative, and a more relaxed mother. The marital relationship has also improved. They are more supportive of each other and are more able to communicate while controlling some of their arguments. This last accomplishment is one of which they are both extremely proud. Again, referring to the time line (Figure 2), it can be noted that Jimmy's hospitalizations and sick visits increased during the period of highest marital stress and decreased after the marital relationship improved.

DR. KENNETH G. REEB (Associate Professor, Department of Family Medicine): Today's presentation demonstrates the utility of the concept of three levels of family care: "thinking family," "working with families," and "doing family counseling." Ms. Levitt has been involved in all three of these levels. In "thinking family," she has considered family implications of chronic disease as she and her instructor have provided ongoing general health care to all family members. They have managed complications of the child's sickle cell disease, such as vaso-occlusive crises and fever, with consultation from the pediatric hematologist.

The book, The Family Is the Patient, 1 by Allmond et al has an excellent chapter about families of children with chronic disease. It urges physicians interested in "working with families" to arrange a family conference. Standard medical practice involves interaction between the physician, the sick child, and the mother, but rarely is the father or the family as a whole involved. A family conference allows the clinician to make clinical observations of family and individual problems. For example, a common pattern of families who have a child with chronic disease includes a mother who is overinvolved with her baby and a father who is forced to work extra hours to help finance the tremendous costs of the chronic dis-



ease. His absences also allow him to repress his fears about his sick baby and to avoid getting jealous of his wife's preoccupation with the baby. Family physicians can use a scheduled family conference to detect these maladjustive patterns.

A developmental family life cycle concept can help explain the family's reaction to chronic disease. LeMasters proposed the concept of "childbirth as a crisis." The birth of a sick infant during this crisis predisposes to a poor, rather than a good, adjustment to the task of becoming parents. Parenthood can be viewed as a developmental process for parents as well as for children. Benedek says, "When the parent consciously tries to help the child achieve his developmental goals, he cannot help but deal with his own conflicts consciously and by these, normally achieve a new level of maturation."3 Children with sickle cell disease experience difficulties in attaining the developmental tasks of early infancy, especially the task of developing basic trust.4 Sickle cell anemia deprives parents of the opportunity to relive a successful, early stage of development with their baby and, by so doing, to improve their own sense of basic trust. The concept of psychosocial transmission within families also applies to this family. Both parents have major problems with feelings of inferiority. Their child is at risk to have similar problems. This risk is compounded by his disease. Clinicians should take every opportunity to facilitate self-concept development in the child as well as in the parent.

A chronic condition such as sickle cell disease also interferes with the development of the siblings. In this family, the parents are already comparing their unaffected child with the patient. They say they worry less about their daughter, implying that their chronically ill son gets more of their attention. Dr. Medalie's concept of the "hidden patient" applies to siblings as well as to caretakers of the sick patient. All three of the nonchronically ill members of this family are "hidden patients." The family physician should help direct the family's attention back to the siblings to help them deal with their adjustment problems.

MRS. ANTONNETTE V. GRAHAM (Senior instructor, Department of Family Medicine): In caring for the B. family, Ms. Levitt was able, as Dr. Reeb discussed, to be involved with the family at the three levels of "thinking family," "working with the family," and "doing family counseling."

Ms. Levitt was able to provide "family counseling" for several important reasons, the most important of which was the relationship she has already established with the family. As Geyman⁵ and Castelnuovo-Tedesco6 have noted, the family physician is an ideal health professional to offer counseling to patients. Schmidt and Messner⁷ and Martin⁸ write specifically about the family physician as being a marital counselor. The family physician is able to become helpful to a troubled family because of the trusting relationship that has already been established between the family and the physician. This was very evident in the work Ms. Levitt did with the B. family, especially with Mrs. B. Although Mrs. B. had been known to a wide variety of health professionals through her own extended and her son's numerous hospitalizations, she had difficulty forming relationships and was thought of as a "very difficult patient" by hospital personnel. It was different with Ms. Levitt. Mrs. B. turned to her student physician as someone she could trust and saw Ms. Levitt as someone who would help her. This relationship then became the basis for the subsequent marital counseling. Here in the Department of Family Medicine at Case Western Reserve University, we have found in our educational program with residents that families readily accept their physicians as marital counselors and are very pleased when their physicians want to help them in this very troubling area of their life.9

In doing marital work, one of the areas most often needing intervention is the communication between the couple. When Ms. Levitt and Mr. Fine began working with them, their communication was suffering. The first videoclip shown during this Grand Rounds pointed this out graphically. Mr. and Mrs. B. sat turned away from each other and spoke mainly to their student physician. They both noted that they could no longer "talk to each other." One of the goals for counseling was not only to improve the couple's communication but to help them discuss together Jimmy's illness. The impact of a child's serious illness on a family can be devastating. When families are not able to accept and deal with their child's illness, serious implications for the family occur, often resulting in marital separation or divorce. As Ms. Levitt and Mr. Fine worked with the B. family, their communication patterns improved, and on the last video session shown today, we saw Mr. and Mrs. B.

discussing their feelings and their fears about Jimmy's illness. Mr. B. talks about how the sickle cell disease "plays with them"; sometimes Jimmy is well and Mr. B. dares to "hope," but then a crisis occurs and Mr. B. fears for his son's life. Mrs. B. quietly tells that she wishes she could bear the pain instead of her little boy. Each agreed that talking about their son's illness was difficult, but necessary, so that they did not suffer alone.

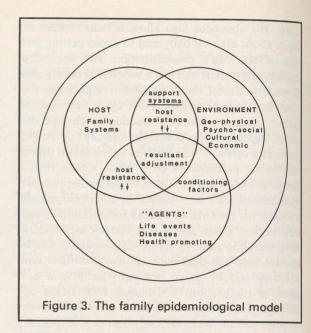
As the couple began to relate more positively to each other, and to deal more openly with Jimmy's illness, fate intervened in a very positive way. Darla, a normal, healthy child was born. The parents were able to relate to her normalcy as a reinforcement of their ability to do something good together. Successful marital treatment depends upon several factors, two of which are supportiveness of both treatment and extra therapeutic (home, work, community) environment. 10 During the period from December 1980 to March 1981. Jimmy's hospitalizations and sick visits to the Family Practice Center decreased and his general health improved. Here we can see the interrelationship of the B.'s improved marital relationship and Jimmy's improved health status.

Finally, I would like to briefly address the educational experience for Ms. Levitt and Mr. Fine. At no time were our goals to make them marriage therapists in the traditional sense of the word. With close supervision, review of videotapes, and numerous readings, we hoped to give them skills that would be useful in their future roles as primary care physicians.

DR. JACK H. MEDALIE (Professor and Chairman, Department of Family Medicine): What we have seen and heard today has been most heartening and confirms our feelings that certain students, with appropriate supervision, can become effective counselors and an important support system for the family.

We have also heard a discussion of a number of important concepts of family work. Let me list them:

- 1. Levels of family care
- 2. Family as the patient
- 3. Hidden patient—in this case the family (overlap with No. 2)
- 4. Developmental life cycle concept emphasizing the growth and development of the parents as the child grows
- 5. Psychosocial transmission in the family¹¹



- 6. The relationship of marital satisfaction to the health of the child
- 7. Some principles of marital counseling To this list let me add that I think a good way of looking at this family would be by using the family epidemiological model¹² (Figure 3).

I would like to add a few points to the discussion. The first is that young children with sickle cell disease have a characteristic, distinctive cry which is presumably related to pain, even though we cannot always find objective signs of the cause. A few years ago I worked for a short time with a nurse in rural North Carolina who told me she had no difficulty in recognizing a "sickle cry."

Using the developmental model that Dr. Reeb brought up, I would stress that the pregnancy period is a most appropriate time to help the woman and her husband. For reasons which are probably related to physiological changes, the parents are very open to help during this period and respond very well.

Concurring with Dr. Reeb again, the hidden patient in this instance is definitely the whole family, and the support given by Ms. Levitt and Mr. Fine came at a vital time in this family's development. Regarding their support systems, it is most important for us to understand the major role played by the church for this family, and a meeting between

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SLOW-RELEASE TABLETS, 10 mEq

DESCRIPTION KLOTRIX is a film-coated (not enteric-coated) tablet containing 750 mg potassium chloride (equivalent to 10 mEq) in a wax matrix. This formulation is intended to provide a controlled release of potassium from the matrix to minimize the likelihood of producing high localized concentrations of potassium within the gastrointestinal tract

INDICATIONS—BECAUSE OF REPORTS OF INTESTINAL AND GASTRIC ULCERATION AND BLEEDING WITH SLOW-RELEASE POTASSIUM CHLORIDE PREPARATIONS, THESE DRUGS SHOULD BE RESERVED FOR THOSE PATIENTS WHO CANNOT TOLERATE OR REFUSE TO TAKE LIQUID OR EFFERVESCENT POTASSIUM PREPARATIONS OR FOR PATIENTS IN WHOM THERE IS A PROBLEM OF COMPLIANCE WITH THESE PREPARATIONS.

1. For therapeutic use in patients with hypokalemia with or without metabolic alkalosis: in digitalis intoxication and in patients with hypokalemic familial periodic paralysis

2. For prevention of potassium depletion when the dietary intake of potassium is inadequate in the following conditions: Patients receiving digitalis and diuretics for congestive heart failure; hepatic cirrhosis with ascites; states of aldosterone excess with normal renal function; potassium-losing nephropathy, and certain diarrheal states

3. The use of potassium salts in patients receiving diuretics for uncomplicated essential hypertension is often unnecessary when such patients have a normal dietary pattern. Serum potassium should be checked periodically, however, and, if hypokalemia occurs, dietary supplementation with potassium-containing foods may be adequate to control milder cases. In more severe cases

supplementation with potassium salts may be indicated.

CONTRAINDICATIONS In patients with hyperkalemia, since a further increase in serum potassium concentration in such patients can produce cardiac arrest. Hyperkalemia may complicate any of the following conditions: chronic renal failure, systemic acidosis such as diabetic acidosis, acute dehydration, extensive tissue breakdown as in severe burns, adrenal insufficiency, or the administration of a potassium-sparing diuretic (eg, spironolactone, triamterene)

Wax-matrix potassium chloride preparations have produced esophageal ulceration in certain cardiac patients with esophageal compression due to enlarged left atrium

All solid dosage forms of potassium supplements are contraindicated in any patient in whom there is cause for arrest or delay in tablet passage through the G.I. tract. In these instances, potassium supplementation should be with a liquid preparation

WARNINGS Hyperkalemia: In patients with impaired mechanisms for excreting potassium, administration of potassium salts can produce hyperkalemia and cardiac arrest. This occurs most commonly in patients given potassium intravenously but may also occur when given orally. Potentially fatal hyperkalemia can develop rapidly and be asymptomatic. Use of potassium salts in patients with chronic renal disease, or any other condition which impairs potassium excretion requires particularly careful monitoring of the serum potassium concentration and appropriate dosage adjustment.

Interaction with potassium-sparing diuretics: Hypokalemia should not be treated by the concomitant administration of potassium salts and a potassium-sparing diuretic (eg, spironolactone or triamterene), since the simultaneous administration of these agents can produce

severe hyperkalemia

Gastrointestinal lesions: Potassium chloride tablets have produced stenotic and/or ulcerative lesions of the small bowel and deaths. These lesions are caused by a high localized concentration of potassium ion in the region of a rapidly dissolving tablet, which injures the bowel wall and thereby produces obstruction, hemorrhage, or perforation. KLOTRIX is a wax-matrix tablet formulated to provide a controlled rate of release of potassium chloride and thus to minimize the possibility of a high local concentration of potassium ion near the bowel wall. While the reported frequency of small-bowel lesions is much less with wax-matrix tablets (less than one per 100,000 patient-years) than with enteric-coated potassium chloride tablets (40-50 per 100,000 patient-years) cases associated with wax-matrix tablets have been reported both in foreign countries and in the United States. In addition, perhaps because the wax-matrix preparations are not enteric-coated and release potassium in the stomach, there have been reports of upper gastrointestinal bleeding associated with these products. The total number of gastrointestinal lesions remains less than one per 100,000 patient-years. KLOTRIX should be discontinued immediately and the possibility of bowel obstruction or perforation considered if severe vomiting, abdominal pain, distention, or gastrointestinal bleeding occurs.

Metabolic acidosis: Hypokalemia in patients with metabolic acidosis should be treated with an alkalinizing potassium salt such as potassium bicarbonate, potassium citrate, or potassium acetate. PRECAUTIONS Potassium depletion is ordinarily diagnosed by demonstrating hypokalemia in a patient with a clinical history suggesting some cause for potassium depletion. In interpreting the serum potassium level, the physician should bear in mind that acute alkalosis per se can produce hypokalemia in the absence of a deficit in total body potassium, while acute acidosis per se can increase the serum potassium concentration into the normal range even in the presence of a reduced total body potassium. Treatment of potassium depletion particularly in presence of cardiac disease, renal disease, or acidosis, requires careful attention to acid-base balance and appropriate monitoring of serum electrolytes, electrocardiogram and clinical status of patient. ADVERSE REACTIONS Most common to oral potassium salts: nausea, vomiting, abdominal

discomfort, and diarrhea. These symptoms are due to irritation of the gastrointestinal tract and are best managed by diluting the preparation further, taking the dose with meals, or reducing the dose. One of the most severe adverse effects is hyperkalemia (see Contraindications and Warnings) There also have been reports of upper and lower gastrointestinal conditions including obstruction, bleeding, ulceration and perforation (see Contraindications and Warnings); other factors known to be associated with such conditions were present in many of these patients. Skin rash has been reported rarely

DOSAGE AND ADMINISTRATION The usual dietary intake of potassium by the average adult is 40 to 80 mEq per day. Potassium depletion sufficient to cause hypokalemia usually requires the loss of 200 or more mEq of potassium from the total body store. Dosage must be adjusted to the individual needs of each patient but is typically in the range of 20 mEq per day for the prevention of hypokalemia to 40-100 mEq per day or more for the treatment of potassium depletion.

Note: KLOTRIX® slow-release tablets must be swallowed whole and never crushed or chewed. Following release of the potassium chloride, the expended wax matrix, which is not absorbed, may

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FAMILY AND SICKLE CELL ANEMIA

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the minister and the family practice team would not be out of place.

A child with congenital chronic disease, particularly the firstborn, places a tremendous strain on the family, and this family illustrates how important the birth of a second normal child is. The child is a tremendous boost for the parents' self-esteem. and Darla's "normality" becomes part of the support system as long as they do not split into two dyads-the mother and Darla against the father and Jimmy.

Finally, I think it would be important for the parents to become involved with other parents of sickle cell children if there are any "self-help" or "sickle cell anonymous" groups in the community.

For the future, I believe it will not be long before the appropriate prenatal diagnosis of sickle cell anemia by amniocentesis will be available; then there will be an opportunity to prevent the birth of affected children. However, this will involve ethical and religious issues, so sickle cell disease might be with us for a long time.

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