Family Practice Grand Rounds

An Adolescent Looking at Chronic Illness

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DR. HARLEY J. RACER *(Program Director, Family Practice Residency):* I have the mixed pleasure of introducing to Grand Rounds today the family practice resident who has displaced me as family physician for this patient and her family.

It has become clear to me that the processes of continuity and comprehensiveness of care which all of us feel are fundamental to family practice can be experienced by the family practice resident during his tenure in the residency program, with today's case presentation serving as an outstanding example. Dr. Clarke Tungseth, our chief participant in today's Grand Rounds can not only discuss our patient's serious and complex presenting problem, systemic lupus erythematosus,[1-3] in depth and in breadth, but also demonstrates an understanding of the dynamics of the family surrounding this patient and how they affect her manner of dealing with her illness.

DR. CLARKE TUNGSETH *(family practice resident):* Our patient today is a 17-year-old woman, whose progressive development of symptoms of arthritis, vasculitis, and nephritis[4,5] over a two-year period led to the recognition of systemic lupus erythematosus. Our intent in Grand Rounds today is to review the disease process of lupus, with an emphasis on recognition[6] and management, and also to explore the impact of such a serious chronic disorder on the life of an adolescent patient, her family and her fiancé.

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Over the past two years I have become the physician to Sherrie's family. Since I first saw her grandmother with an otitis media, my many contacts with members of the extended family have enlarged my understanding of each individual and of the resources and functional capacity of the family as a whole. This has led to a better understanding of the psychological and personality resources with which Sherrie meets her current illness (Table 1).

Sherrie's clinical problem list (Table 2) helps display the interrelationship of biological, psychological, and social issues affecting her care. The chronological flow sheet of her office visits (Table 3) unfolds the natural history of the development of systemic lupus erythematosus which Dr. John Flinn, our rheumatology consultant, will discuss.

DR. JOHN FLINN *(Internist and Rheumatologist):* Let me begin by showing you a preliminary list of fourteen criteria which have been proposed by the American Rheumatism Association as being significant in the recognition of systemic lupus erythematosus (Table 4).[7,8] If four of these criteria are met by the patient, the diagnosis of systemic lupus erythematosus is quite certain in 85 percent of the cases. If five of these criteria are present, the certainty of diagnosis approaches 95 percent, and if more than five of these criteria are present, almost every patient must be considered to have systemic lupus erythematosus.

Let us look at Sherrie (Table 3). In June 1971 she presented with epistaxis (possibly small-vessel disease already!), definite synovitis (a hot, red, warm joint), and a malar rash. The diagnosis of systemic lupus erythematosus was properly considered even at that time, and when she returned just a month later some initial laboratory evaluation for lupus was actually begun. But then we see one of the real pitfalls of the developing diagnosis of a disease like lupus with an insidious onset: *when the initial sedimentation rate and anti-DNA were not positive, concern about lupus was set aside,* even though two of the clinical criteria were already met! A third criterion, photosensitivity, was met in May 1972, ten months later, and a fourth criterion, pleurisy, occurred 14 months after onset. The fifth criterion was undoubtedly met by the fainting episode in August 1973, 26 months after the first signal of her illness, but it was not until the onset of the nephrotic syndrome in December 1973, 30 months after onset, that the disease condition was fully recognized.

Sherrie's experience is not unusual, and the lesson we need to learn is that we must return to our basic responsibility as clinicians who recognize illness with old-fashioned history and physical skills, which yield trustworthy information. We must be able to interpret this unfolding story if we are to recognize the progressing vasculitis of this serious condition, signified by symptoms occurring in several systems.

By being well informed, both from the patient and about the disease, we should be able to recognize systemic lupus erythematosus and institute appropriate therapy much earlier from now on.

Systemic Lupus Erythematosus: Treatment

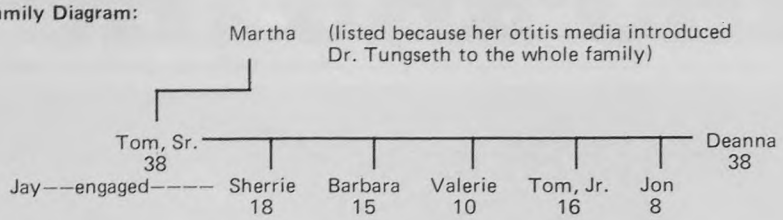
DR. TUNGSETH: By looking at the flow sheet on Sherrie's visits (Table 3) you can see the progress of her steroid therapy and its complications, the regression of her lupus nephritis, and now the introduction of a serious clinical problem, that of pregnancy in the lupus patient.

Before we proceed with some further background regarding Sherrie's family, psychological, emotional, and social situation, we will return to Dr. Flinn for a discussion of the management of the underlying illness.

DR. FLINN: In the overall management of the patient with systemic lupus erythematosus, patient education is the very foundation of cooperative management by the patient and the doctor of the many complications produced by vasculitis which can affect any organ or system. The

Table 1. Family Situation

A. Family Diagram:



B. Family Profile:

Tom, Sr. —High school teacher (mathematics), golf coach, drivers' education, bus driver. Stable, head of the family. Generally good physical and emotional health.

Deanna —Quite busy raising five children and supporting her husband. Tends to react to the strain and has frequent minor complaints, tiredness, etc. Also brings children in with minor complaints. Worries too much.

Sherrie —High school graduate. Trying different jobs at this point. Engaged. Warm, enjoyable.

Barbara —Healthy and without problems until diagnosis by me of fairly marked idiopathic scoliosis 1/74. Is responding well to treatment with Milwaukee Brace. Has been in once with anxiety and discouragement related to wearing brace.

Valerie, Jon, Tom, Jr. —Good general health.

Family lives in own home, spends much of summer in lake cabin. Seems to be close and relate well.
Family historically has used medical facilities frequently.

Jay (fiancé) —Mother died of cancer when Jay was in elementary school. Father died soon after with liver disease secondary to alcoholism. Jay lived with an older brother, Will (now in marital trouble) and a sister, Carrie. Jay is described as nice, understanding, but does not manage money wisely.

Table 2. Problem List

Problem #1	Systemic lupus erythematosus	Onset: June 1971
	a. Lupus glomerulonephritis	Onset: December 1973
	b. Chronic cortisone therapy	Onset: January 1974
Problem #2	Pregnancy	Onset: November 1974
	a. Risks associated with lupus erythematosus	
	b. Future contraceptive planning	
Problem #3	Life situation	
	a. Independent/dependent	Onset: January 1974
	b. Marriage decision	Onset: 1972
Problem #4	Reactive depression	Onset: December 1973

patient must have the best possible understanding of her question, "What's the matter with me, Doctor?"

The answer, ". . . systemic lupus erythematosus, a vascular disorder which causes disturbances of function in many organs and systems . . ."[10] is only a beginning of the explanation and the understanding. In a chronic disease of this magnitude and extent of dissemination, and with such serious implications for critical malfunction, the patient is apt to shop far and wide for magic answers unless rapport is established on the basis of clear understanding. From then on, the treatment of the minor manifestations, the arthritis, the skin lesions, the febrile episodes, and those manifestations which do not involve major organs (kidneys, heart, lungs) is much more readily achieved. The patient will be able to cooperate sensibly with the use of minor anti-inflammatory agents such as aspirin, indomethacin, or even chloroquine (when there is considerable skin involvement). Steroids[11] are, of course, reserved for major system involvement, and occasionally to quiet more disabling joint inflammation in order to maintain a satisfactory quality of life.[12]

Of course, the employment of the steroids introduces all of those complications we see Sherrie experiencing (Table 3), and requires close supervision to recognize gastritis, hypertension, and serious psychological disturbances that might be related to steroid therapy.

Close observation during periodic exacerbation of symptoms and during steroid therapy is tailored to the severity of symptoms or side effects, but even when the disease is relatively quiescent, monthly visits are required for up to six months, and thereafter at two to three-month intervals for maintenance observation.

Some of the deeper questions that must be considered with the patient are those that concern potential disability, productivity, marriage, and the very serious concern about pregnancy.[7] Pregnancy for the lupus patient represents a serious physiologic stress for the individual with widespread vasculitis. Exacerbations of major organ disorders in kidneys, heart, and lungs are common during pregnancy, delivery, and the puerperal recovery period. Many physicians consider pregnancies such a threat to the lupus patient, with such serious potential for further major organ damage, that therapeutic abortion has been considered justified in order to reduce major organ injury due to exacerbation of lupus during pregnancy. The decision to recommend therapeutic abortion, however, is not simple, since in some patients who have undergone therapeutic abortion, exacerbations of their vasculitis have been even more severe than in those patients who have completed pregnancy and uncomplicated delivery. It is a decision that will have to be entered into on an individual basis, bearing in mind the severity of the patient's active vasculitis, her life situation, and the value and meaning of pregnancy for her in the face of its added risks.[7]

Life Situation

DR. TUNGSETH: Now I want to show you three small video-tape segments of interviews with Sherrie, her father, and her fiancé, that I think will demonstrate some of the individual and family dynamics, and the relationship issues that are involved so deeply in this serious illness. (Shows a videotaped interview with Sherrie's father):

DR. TUNGSETH: "Tom, I'd like to know how you, your wife Deanna, and the rest of the family have reacted to Sherrie's being sick. I've been concerned especially about this since you've had to absorb this news and the

problem that her sister Barbie has had to go into a brace to correct her scoliosis. How are you all feeling about these problems?"

TOM, SR.: "When we first heard about Sherrie, I could tell that Deanna was really very uptight about it, so I told her that there just wasn't anything right now that we had to get so worked up about. I said, I'm going to look on it from the standpoint that it's just nothing very serious. I said you could make it just as serious as you want to, knowing nothing about it, you could make it so serious that she's going to be dead in a month. And I said, until we get the report or until the doctor gets through with her, it's certainly going to bother us, but the rest of the family has to live, the rest of us have to get going on.

Then Barbie gets this (scoliosis)! And, of course, when we find out that it's nothing from the standpoint of dying, in both cases, why then it's practically like all the pressure is off."

DR. TUNGSETH: Now I'd like to show you a brief segment of a number of interviews that I've had with Sherrie herself about her illness and her view of her situation as a whole. (Shows video-tape segment of interview with Sherrie):

SHERRIE: ". . . and to hang curtains and paint and that kind of thing. I like to do those kinds of things but Jay really doesn't. That's one of the things that was really hardest about the lupus because one time I had these shades in my room that rolled out and came out of the wall and they fell on me. I tried to put them back up again, but I was so fat that I couldn't move as well as I did before. My bed is very high and I fell off my bed and the curtains came falling down and I took the curtain and threw it across the room and ran upstairs and said, 'You go down and fix that curtain for me because I can't do it now.' I was really upset because I couldn't do it. I've always been the kind of person who moves her own bedroom furniture around even though I do have a king-size bed and that king-size bed is hard to move. I've adjusted to it."

DR. TUNGSETH: "You mean you've learned to take it, is that what you mean adjusting?"

SHERRIE: "Uh huh, anything I can't do, I ask for help but usually I try to see if I can do it and if I can't . . ."

Table 3. Chronological List of Office Visits

1/69	Upper respiratory infection. First clinic visit.
6/71	Headache (school conflicts – performance). Recurrent epistaxis, nevus, dysmenorrhea, synovitis right knee, rash across cheeks and nose.
7/71	Recurrent facial rash. Sedimentation rate – 23 Anti-deoxyribonucleic acid nuclear antibody – negative.
3/72	Epistaxis.
5/72	Rash on forearms and ankles. Sun vs fertilizer on grass.
10/72	Intermittent chest and abdominal pain. Exam negative.
3/73	Breast lump. Headache (school problems – friends).
8/73	Fainting episode vs seizures. Exam normal. Electroencephalogram normal. (My first contact).
12/73	Headache, dysmenorrhea, ankle swelling in morning. Proximal interphalangeal stiffness. Exam normal. Blood pressure 110/70. Urinalysis: 2+ albumin. Sedimentation rate – 70.
12/73	Recheck. Albumin – 3+. Strep screen – negative. Antistreptolysin – negative.
12/73	Recheck nocturia one time: albumin – 3+. Serum albumin – 2.8. Globulin – 2.8. 24-hour urine: protein 8.7 grams, creatinine – .8, cholesterol – 345, sedimentation rate – 88.
1/74	Admit. Kidney biopsy. Diffuse epimembranous glomerulonephritis. Glomerular deposits, complement$_3$, immunoglobulin G. Lupus erythematosus clot negative. Rheumatoid factor negative. Fluorescent antinuclear antibody: 1:10. Complement$_3$, complement$_4$ – normal. Coombs' – negative. Hemoglobin – 11.6. Albumin – 1.8, creatinine clearance – 119.
1/74	Fluorescent antinuclear antibody: 1:80. Start prednisone 60 mg daily.
1/74	Weight gain, edema. Treated with triamterene (Dyazide).
1/74	Recheck. Edema gone. (Fiancé leaves for military duty).
1/74	Urine: albumin 1+. Return to school. Albumin 3+, joints flare. Treatment – more rest.
2/74	Improved but moon face, buffalo hump, thinning of skin, mild acne, interference with sleep, central weight gain, and appetite problems. Albumin 1+, serum albumin 3.7, hemoglobin – 15.6, sedimentation rate – 31. Start tapering prednisone (now two months, 60 mg daily).
3/74	Urine – albumin trace.
4/74	Upper respiratory infection.
4/74	Urine – albumin negative. Prednisone 30 mg every other day. Malar rash with sun. Pleuritic chest pain. Amenorrhea.
5/74	Cystitis. Urine – albumin negative.
6/74	Sun exposure – rash, malar and forearm. Treatment: continue same prednisone 20 mg every other day, aspirin, sun screen, and avoidance.
6/74	Rash cleared. Urine – albumin 4.7. Sedimentation rate – 21. Prednisone 10 mg every other day. Epigastric distress relieved by antacids. Treatment: antacids, stop aspirin, Darvocets for pain.
7/74	Prednisone 4 mg every other day. Periods May, June, July.
9/74	Episode back pain, blurred vision, faintness. Prenisone 3 mg every other day. Urine – albumin negative. Tension headaches (threatened relationship with Jay).

DR. TUNGSETH: Here now is a brief sample of an interview with her fiancé. (Shows video-tape segment of interview with Jay):

JAY: ". . . it probably made it stronger – the feeling of getting married, because I knew she needed someone to look after her and I was the one as far as I'm concerned. It seems like her sickness brought us a little closer. I couldn't sit and think of the way she was going through pain like this, it just turned my stomach. If I could just help so that she won't feel it again . . ."

Psychological Evaluation

DR. TUNGSETH: Now I would like to introduce Dr. Harry Ireton, my clinical psychology preceptor who saw Sherrie with me, and who will discuss some of the findings of her psychological evaluation.

DR. HAROLD R. IRETON *(Clinical Psychologist):* As the oldest daughter, Sherrie has become the caretaker for others, the person on whom others can rely. She is outgoing, likes people, is very much involved with them, and is oriented to meeting the needs of others. She has developed interests in drama, speech, music, and physical education (especially track). She says she especially likes to compete with boys and win, although she claims not to carry this to extremes, and avoids this with her fiancé, Jay.

In our interview, Sherrie was friendly, presented herself as cheerful and optimistic, but appeared somewhat tense and sad. Sherrie places a high value on "strength, independence, perseverance, and activity." She is very much a doer. She is a very caring person, who is oriented to meeting the needs of others, often at the expense of her own. In other words, she finds her value in doing for others. Feelings of loss, disappointment, depression, or anger are incompatible with her life-

Table 4. Preliminary Criteria for Classification of SLE (14 manifestations)[7,9]

Facial erythema
Discoid lupus – face and any other skin area
Raynaud's phenomenon
Alopecia
Photosensitivity
Oral or nasopharyngeal ulceration
Arthritis without deformity
Lupus erythematosus cells
False positive serologic test for syphilis with negative treponema pallidum immobilization
Profuse proteinuria (<73.5 gm/day)
Cellular casts
Pleuritis or pericarditis
Psychosis or convulsions
Hemolytic anemia
Leukopenia
Thrombocytopenia

Four or more presenting serially or simultaneously during any interval of observation = positive diagnosis of lupus erythematosus.

style, and so are managed by suppression and denial.

Regarding her illness, she expresses concern that she "will be left out of things, on the outside looking in." She tends to maintain her physical activities, even when this is unrealistic because of present symptoms, so that others will not be so concerned about her. She is very reluctant to depend on others, emotionally or otherwise, because this might jeopardize her need to see herself as "strong and self-sufficient." Even while ill, she is more concerned about others than herself. Her underlying sense of loss and depression, and probable anger, have not been faced by Sherrie.

There are four major psychological issues which we must deal with in the case of Sherrie: (1) decisions regarding pregnancy [most pressing]; (2) counseling for Sherrie to help her to express her feelings and personal needs more, to encourage her to occasionally allow herself to use others as a source of emotional support, and to support her values of being active and caring for others, but on a more realistic basis; (3) counseling for the family regarding an understanding of the illness and the psychological issues; and (4) occupational activity for Sherrie.

Sherrie has literally to be given permission to acknowledge her feelings and her needs. She has to be gently encouraged to allow herself to occasionally lean on another human being for emotional support; and while the counselor or physician is doing that (and it is a balanced sort of thing), he must also acknowledge her values and the importance of activity to her. We cannot take that away from her. We have got to help her do that on a more realistic basis. She is basically a tremendous human being. Now the underside of that coin is that family and society often pay people (physicians are a good example) for killing themselves, so that she has been tremendously reinforced for playing this particular role. In terms of the family, we should meet with them to get more of a sense of how they see these things. If we could help Sherrie to function in some relevant way occupationally, it would be very important to her to be active in that regard.

DR. TUNGSETH: In summary, we have reviewed the medical and surrounding psychological and social issues of a young woman facing a chronic illness. We have traced the developing natural history of a syndrome of synovitis, photosensitivity dermatitis, pleuritic pain, apparent seizures, and nephrotic syndrome, all leading to the establishment of the diagnosis of systemic lupus erythematosus. The disease was followed through a course of therapy with intensive corticosteroids and its attendant side effects of moon face, acne, weakness, central obesity, buffalo hump, thinning of the skin, gastric irritation, and amenorrhea. The therapeutic response, the tapering of the corticosteroid medication, and gradual return to normal activity have been described. The medical aspects of this case have been considered in the context of the patient's personality, and her surrounding psychosocial considerations. The patient's unnecessarily brave front, attempting to protect her family and fiancé from her suffering, has been noted. The patient's health team has tried to help her address her emotional needs in the context of making decisions about her pregnancy and the possibilities of marriage. The patient has found this comprehensive approach useful, and has been able to deal realistically with her disease and her feelings and to feel confident in making her own decisions.

DR. RACER: What we have seen today is a Family Medicine Conference[13] produced and conducted by the family physician as one instrument for the integrated teaching of family medicine. It is the clinical conference in which the family physician presents common, serious, and model problems which he meets and deals with in both health and illness, drawing on resources and specialists from many disciplines to present the balanced integration of behavioral and organic elements that must be considered in caring for and in caring about our patients.

Addendum

Although advised that the risk of continuing the pregnancy with her illness was acceptable, Sherrie did elect to terminate the pregnancy. Later Sherrie and Jay were married. They are both working and have been doing well. The lupus erythematosus has remained in remission on low-dose, alternate-day prednisone.

References

1. American Medical Association: Primer on rheumatic diseases. Section 8: Systemic lupus erythematosus. JAMA 224:701-711, 1973
2. Keefer CS, Wilkins RW: Medicine Essentials of Clinical Practice. Boston, Little, Brown, 1970
3. McClary AR, Meyer E, Weitzman EL: Observations on the role of the mechanism of depression in some patients with disseminated lupus erythematosus. Psychosom Med 17:311-321, 1955
4. Decker JL, Steinberg AD, Gershwin ME, et al: Systemic lupus erythematosus: Contrasts and comparisons. Ann Intern Med 82:391-404, 1975
5. Baldwin DS, Lowenstein J, Rothfield NF, et al: The clinical course of the proliferative and membranous forms of lupus nephritis. Ann Intern Med 73:929-942, 1970
6. Fraga A, Mintz G, Orozco J, et al: Sterility and fertility rates, fetal wastage and maternal morbidity in systemic lupus erythematosus. J Rheumatol 1:293-298, 1974
7. Cohen AS, Reynolds WE, Franklin EC, et al: Preliminary criteria for the classification of systemic lupus erythematosus. Bull Rheum Dis 21:643-648, 1971
8. Ginzler EM, Nicastri D, Chen C, et al: Progression of mesangial and focal to diffuse lupus nephritis. N Engl J Med 291:693-696, 1974
9. American Medical Association: Appendix: Criteria for diagnosis and classification of rheumatic diseases. JAMA 224:801, 1973
10. Stiller CR, Russell AS, Dossetor JB: Autoimmunity: Present concepts. Ann Intern Med 82:405-410, 1975
11. Yount WJ, Utsinger PD, Puritz EM, et al: Corticosteroid therapy of the collagen vascular disorders. Med Clin North Am 57:1343-1355, 1973
12. St. Louis Park Medical Center Research Foundation: Lupus and You: A Guide for Patients. Minneapolis, St. Louis Park Medical Center, 1972
13. Burr BD: Family practice grand rounds: Teaching behavioral science in family practice. In, Asilomar Conference Proceedings. Davis, California, Department of Family Practice, University of California, Davis, 1973