Epidermolysis Bullosa: A Zinc Dependent Condition?

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In Harrison's book, *Principles of Medicine*, 10th Edition, Petersdorf et al. (1983), an inherited zinc deficiency disease is described. The symptoms and signs include severe chronic diarrhea, muscle wasting, alopecia, rough thick ulcerated skin about the body orifices and on the extremities. Recently I was consulted by a young man and his mother for help in the treatment of his severe intractable case of epidermolysis bulosa. His disease and his response to administration of zinc and a few other nutrients raises the possibility that his condition is a variant of a chronic zinc dependency.

CC came to see me on December 19,1989, and was seen for the second and last time January 5, 1990. A few days after he was born in June 1972 his skin which had been under pressure from a forceps delivery began to slough of. A few days later lesions developed on his face, mouth, chest and limbs, which later blistered. He was treated with topical antibiotics using sterile technique but was not better. The lesions in his mouth made it impossible for him to feed and he became anemic and hypoproteinemic. One month later he was diagnosed epidermolysis bullosa and started on vitamin E 600 LU. orally, later increased to 800 LU. At age 4 months 200 mg of ascorbic acid was added and at age 6 months he was given iron supplementation and the vitamin E was increased to 1000 LU. There was no response. He was then admitted to hospital suffering from stomatitis and again in April 1973 for gastroenteritis and pneumonia. By now he had multiple lesions and denuded areas on his legs, no nails and adhesions between his fingers and toes. On top of these lesions he had been constantly constipated ... His mother had to remove his stools manually daily. In 1986 he was admitted to hospital to have his stools removed. In 1980 his parents took him to West Germany for two and one half months to be treated by a biochemist who was using special skin salves and other treatment, with some success. He was placed upon a vegetarian diet supplemented with moderate vitamin program, doses unknown. This regimen was helpful. They went back to Germany once more for ten days and would have gone again but they could no longer afford to do so.

When I first saw him he appeared to be about ten years old, very short and immature. Mentally he appeared to be normal. There were no perceptual changes, no thought disorder and his mood was surprisingly cheerful and upbeat. The bullous lesions continued to erupt. He had lost all of his fingers and toes. An attempt had been made to separate them surgically in Italy with no success. He did tell me that food did not taste normal. He was still severely constipated. To test his sense of taste for a possible zinc deficiency I gave him a teaspoon of a special zinc sulfate solution. He found it tasted like stale water, not bitter as it would to normal individuals. I could not order a blood test for zinc since all his superficial veins were gone and it would have required a cut down. Zinc deficiency will cause Dwarfism, retarded wound healing and loss of taste. The classic response to chronic zinc deficiency is acrodermatitis enteropathica (a.e.). AE babies develop infections around their body orifices (Petersdorf et al., 1983; Prasad, 1976).

I advised him to start on the following supplements: niacinamide 500 mg bid, ascorbic acid 500 mg tid, Pyridoxine 100 mg bid, cod liver oil 1/2 teaspoon daily, ten drops bid of a solution of zinc sulfate 10% with manganese chloride 1/2% plus 1 teaspoon of linseed oil daily to increase his intake of omega three essential fatty acids.

Two weeks later he was much improved. His mood remained normal but his parents were much more cheerful. In that brief period he had grown 1/2 an inch in height, his skin was much healthier and the lesions came on about

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one third as frequently. Those that did develop healed much more quickly. He had gained two pounds. He was no longer constipated and was able to have normal bowel movements for the first time in his life.

When I first saw him his mother informed me that he was very sensitive to any tablets. I therefore had him start one nutrient at a time and if there was no bad reaction he was to proceded to the next. He had no bad reactions to any of the supplements. I increased his niacinamide to 500 mg tid and the ascorbid acid to 1 gram bid.

I have already referred to Harrison's book where chronic zinc deficiency is described. Prasad (1975) also provides good clinical descriptions of the effect of zinc deficiency. This patient's rapid response does not prove it was due entirely to the administration of the zinc. I suspect that zinc was the main therapeutic variable but the other nutrients must also have played an important part. Single nutrient deficiencies are very rare ... I could not in good conscience withhold the other nutrients since I was not certain that one alone might help. This can be determined later on by withdrawing one at a time to determine which are the most important. This family had already suffered too much and needed relief as quickly as possible, nor could they have stayed in Victoria any longer.

In my opinion it is very important that doctors who see these cases try this nutrient treatment approach with zinc the main variable but with the use of other supplements as well.

A nutrient deficiency exists when a person's normal requirements are not met by the diet. Thus if a person needs 20 mg of vitamin B_3 per day to remain free of pellagra and if the diet provides only 10 mg, he will develop
pellagra. If however an individual needs 50 mg per day no known unsupplemented diet will provide that much and he will also develop pellagra. The problem is in the individual, not with the diet, and he has a vitamin B_3 dependency. More is required to maintain health. The amount needed may be very large. For example fifty years ago it was shown that chronic pellagrins required 600 mg per day to remain free of pellagra compared to acute cases who needed much less.

This patient is probably zinc dependent although it is possible he has a multiple nutrient dependency.

October 17, 1991 I spoke to his pediatrician. She had just seen him a week before. She told me that his skin condition was stable, but that he had started to show emotional problems as he matured. This was not surprising since he remains severely handicapped, the aftermath of his severe disease and he will require a good deal of support to cope with the psychosocial problems facing him. On October 24, 1991 this patient called me. He said that his skin condition was stable even though he had remained on the total nutrient approach for one year. Since then he has had minor skin eruptions in the spring and in the fall. He had grown about 3 to 4 inches and had gained 15 pounds. He was still constipated. His mood was level and he was cheerful. He added that he eventually hoped to move to Victoria.

Literature Cited

1. Petersdorf RG et al: *Harrisons Principles of Internal Medicine*, 10th Ed., McGraw Hill, New York, N.Y., 1983.
2. Prasad AS: *Trace Elements in Human Health and Disease,* Vol. 1. Zinc and Copper. Academic Press, New York, N.Y., 1976.