they are likely to be single-gene diseases. Low serum DNase activity in these patients would be consistent with the idea that factors other than \textit{DNASE I} mutations alone affect serum DNase activity, since the odds of an individual inheriting 2 rare gene mutations would be exceedingly low. Four such patients (3 complete C4 and 1 complete C1r deficient) were studied. The number of patients studied is small, reflecting the very low frequency of homozygous C4 and C1r mutations in the population. The results show that serum DNase activity in patients with SLE due to genetic defects in the classical pathway of complement is not significantly different from that in the usual SLE patients (Figure 1). Both groups had serum DNase activity that was significantly diminished compared with that of healthy controls.

We agree with Dr. Balada and colleagues that defects in the ability to clear extracellular DNA likely contribute to the etiopathogenesis of SLE. While mutations in \textit{DNASE I} could be an occasional, but rare, cause of SLE in humans, the etiology of the low serum DNase activity in SLE is undoubtedly complex. Thus far, the data suggest that in most SLE patients this phenomenon cannot be explained solely by \textit{DNASE I} mutations alone. \textit{DNASE I} transcript levels could be influenced by alternative splicing (9) or messenger RNA stability (10) (although there was no evidence of differences in splicing in our earlier study [1]). There is the potential for epistatic interaction with other genes or, alternatively, enzyme activity could be modulated by posttranslational events such as glycosylation, or by microenvironments particular to specific tissues (6,11). Further investigation will be required to determine if any of these processes downstream from gene transcription are affected in SLE patients, or if defects in other endonucleases are present.

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corticosteroids (e.g., prednisone, dexamethasone) in ameliorating asthma (6) or experimental fibrosis in rats (7). It would seem timely to conduct a trial of Lyprinol in a few patients with SSc, perhaps in combination with a steroid.

The mussel from which Lyprinol is derived has been a traditional staple of the diet of the New Zealand Maori people. Advantages of using the lipid extract, Lyprinol, a ×20 mussel concentrate, are that it is prepared from fresh-frozen, stabilized mussels, it is salt-free, and nonallergenic, and it has no solvent residues (liquefied carbon dioxide is being used under supercritical conditions to extract it). There currently are distributors of Lyprinol in the US.

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2. Whitehouse MW, Macrides TA, Kalafatis N, Betts WH, Haynes DR, Broadbent J. Anti-inflammatory activity of a lipid fraction (Lyprinol®) from the NZ green-lipped mussel. Inflammopharma-

DOI 10.1002/art.10343

Congenital fascial dystrophy, a new scleroderma-like genetic disease with limitation of joint mobility: comment on the clinical image presented by Di Rocco

To the Editor:

Congenital fascial dystrophy, described by Di Rocco as stiff skin syndrome (1), is a scleroderma-like syndrome that is virtually unknown to rheumatologists, even though patients with this condition are usually receiving treatment at rheumatology institutions. This autosomal recessive disease is often identified as scleroderm or sclerodermatomyositis because of indurations of the soft tissues, contractures, and limitation of joint mobility. Because of such misidentification, patients may be subjected to aggressive and harmful therapy.

In 1971, Esterly and McKusick (2) described stiff skin syndrome, a condition recognized as a localized connective tissue disorder or limited mucopolysaccharidosis without mucopolysacchariduria. Subsequently, highly heterogeneous disease processes and various scleroderma-like dysmorphic syndromes were reported under this name (3–6). In single cases, Alcian blue deposits were observed between collagen fibers (2,7–9), which favored some relationship with mucopolysaccharidosis.

Our group has reported cases in which patients display stony-hard generalized indurations of the soft tissues, without visceral involvement and no immunologic or vascular abnormalities (10,11). Frequently, these changes are already noticeable on the buttocks and thighs during the first year of life, with progressive involvement of the trunk and limbs. Because of contractures of the limbs, patients have characteristic tiptoe