

Possible Simple Measures for Complex Wound Healing Problems in Ehlers-Danlos Syndrome

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Sir:

Although Hippocrates described the features of Ehlers-Danlos syndrome (EDS), it was named after 2 physicians, viz., Edvard Ehlers from Denmark and Henri-Alexandre Danlos from France, who described it at the turn of the 20th century.¹ It is an inherited disorder of collagen biosynthesis and structure in humans. Joint hypermobility, skin extensibility, abnormal scarring, and tissue friability are the hallmark diagnostic features; however, EDS is under-recognized because when the physical signs are not “classic,” the diagnosis may be elusive. The medical and scientific history of EDS can be seen in 3 phases: clinical characterization, biochemical and molecular genetic analysis, and the use of high-throughput genomic analysis to extend the phenotypes. Over the last years, the characterization of several new EDS variants has broadened insights into the molecular pathogenesis of EDS by implicating genetic defects in the biosynthesis of other extracellular matrix molecules, such as proteoglycans and tenascin-X, or genetic defects in molecules involved in intracellular trafficking, secretion, and assembly of extracellular matrix proteins.² Various types of EDS have been distinguished on clinical and genetic grounds after Villefranche classification, which are not yet incorporated into a coherent classification.

Wound healing is delayed and surgical repair may be difficult because of friable tissues. There is no cure, and treatment is supportive, including close monitoring of the digestive, excretory, and particu-

Table 1. Major Clinical Problems Related to Wound Healing in Patients with EDS and Possible Solutions

Problems of Patients with EDS	Wound Healing Problems Expected in Patients with EDS	Possible Solution(s)	Evidences in the Literature
Defective collagen	Delay in healing and weak scar	Immobilization and rest to healing tissue for prolonged period, particularly during initial stages	More chances of recurrent hernia in patients with EDS ³
Tissue friability	Reduced ability to resist mechanical forces during handling	Gentle handling of tissues during surgery	Malfait and De Paepe ²
	Holding of sutures	Support/immobilization whenever possible	
Skin hyperextensibility	Frequent trauma to healing tissues due to skin mobility and stretching	Delay first dressing change and subsequent less frequent dressing change	Use of negative pressure dressing (LAD) ⁴
		Use of collagen from external source	Use of special bioengineered materials ⁵
		Immobilize part during healing	Faster healing in case reported by Kumar et al ⁴ ; probably because of graft patient is immobilized for longer period and less hindrance to healing tissues occurred

LAD, limited access dressing.

larly the cardiovascular systems. Occupational and physical therapy, bracing, and corrective surgery may help with the frequent injuries.

On searching the literature, we found that few reported encouraging results,³⁻⁵ and we reviewed these reports and tried to find out the reasons for the better

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result. We would like to share possible simple solutions to the problem of wound healing in patients with EDS (Table 1).

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DISCLOSURE

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