pacemaker placement can prolong life in KSS patients with cardiac conduction defects.

Treatments aimed at the primary biochemical defects in mitochondrial encephalomyopathies have been tried; however, the evidence of efficacy has been anecdotal. We generally recommend coenzyme Q10, 50 to 100 mg three times a day, and L-carnitine, 1,000 mg three times a day. Coenzyme Q10 is a quinone compound that normally shuttles electrons from complexes I and II to complex III and may stabilize the oxidative-phosphorylation enzyme complexes within the inner mitochondrial membrane (90). Dichloroacetate inhibits pyruvate dehydrogenase specific kinase, thus activating pyruvate dehydrogenase complex and reducing lactate (91,92). Vitamins that may donate electrons directly to COX include phylloquinone (vitamin K1), menadione (vitamin K3), and ascorbic acid (vitamin C) (54). Vitamin C has also been used as an antioxidant because the impaired oxidative-phosphorylation pathway may generate increased amounts of free radicals. Nicotinamide and riboflavin have been used to improve respiratory chain functions (93). No genetic therapy is presently available.

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