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Management of congenital muscular dystrophies related to defects in the *LMNA* gene

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Congenital Muscular Dystrophies are a heterogeneous group of muscular disorders defined as early onset muscle weakness and progressive course associated to dystrophic features at muscle biopsy. CMDs related to lamina A/C gene (*LMNA*) defect include different phenotypes that can be classified as *i*) severe phenotype with generalized muscular weakness and contractures by birth, *ii*) 'dropped head' phenotype with prominent involvement of axial muscles that generally evolves to rigid spine phenotype and *iii*) early Emery-Dreifuss phenotype. All these conditions generally lead in the first 2 decades to cardiac disturbances, respiratory insufficiency, orthopedic complication and metabolic disorders. The clinical management requires a multidisciplinary and rigorous approach focused on early medical and rehabilitative interventions with the main aims to prevent 'fatal heart event', to cure comorbidities (pulmonary insufficiency and spinal and joint contractures) and to improve the quality of life of these children.

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