

Alpha-Keto Acid Dehydrogenase Complexes (Molecular and Cell Biology Updates)

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Found in all organisms, the alpha-keto acid dehydrogenase complexes have central roles in cellular metabolism and are major sites of regulation. The understanding of the organization, function and regulation of these quintessential multienzyme complexes has been greatly advanced by studies employing molecular biology and biophysical techniques. Although these enzyme systems have some features in common, their diversity in fulfilling unique organism - or tissue - specific roles is truly amazing. These systems have medical importance in areas ranging from defects in regulation (linked to diabetes, heart disease, obesity, nutrition defects), to inherited diseases (inborn errors, maple syrup urine disease) to acquired immune diseases (primary biliary cirrhosis). This book brings together wide-ranging recent findings on the structure(function relationships, gene regulation, and genetic defects of the alpha-keto acid dehydrogenase complexes, namely the pyruvate dehydrogenase, alpha-ketoglutarate dehydrogenase and the branched-chain alpha-keto acid dehydrogenase complexes. A wide variety of experimental approaches together with new results presented in this book should serve as a resource for beginning to established investigators in the field as well as scientists who are interested in mitochondria, dehydrogenases, kinases, phosphatases, lipoic acid, thiamine pyrophosphate, and enzyme complexes.

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