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146. IN VIVO ^{14}C-PROPIONATE INCORPORATION FOR DETECTION OF DEFECTS IN PROPIONATE METABOLISM.

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Propionate metabolic defects may induce propionic acidemia (PA) or methylmalonic acidemia (MMA). Patients with these two inherited automsomal recessive diseases are at high risk of mortality. Confirmatory biomedical diagnosis will help to select appropriate vitamin therapy and genetic conseling. An isotopic method has been established to determine if the propionate metabolic pathway been blocked. The method measures the in vivo incorporation of 1-[^{14}C]-propionate into protein (Trichloroacetic acid, TCA, insoluble material) using cultured cells. The cultured cells were incubated with Puck's saline F containing 1-[^{14}C]-propionate for 18 hrs. After washed with normal saline, 10% TCA was applied to cell layer directly. TCA precipitated materal was then dissolved. The radioactivity and protein concentration of this final reaction mixture was determined by liquid scintillation counting and Lowry's method, respectively. Incorporation rate of propionate in Chinese normal skin fibroblast and normal amniotic cells are established to be 328-2029 pmol/hr/mg protein (n=25) and 184-379 pmol/hr/mg protein (n=20), respectively. The propionate incorporation in an PA skin fibroblast cell lines (GM0371) was determined to be 176 pmol/hr/mg protein by this method. This method provides us a simple and rapid aid of postnatal as well as prenatal diagnosis for the inborn errors of propionate metabolism.