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Retrospective Study

Δ4-3-oxosteroid-5β-reductase deficiency: Responses and long-term outcomes from oral bile acid therapy

Mei-Hong Zhang, Kenneth DR Setchell, Jing Zhao, Jing-Yu Gong, Yi Lu, Jian-She Wang

Abstract

BACKGROUND

Disorders of primary bile acid synthesis may be life-threatening if undiagnosed, or not treated with primary bile acid replacement therapy. To date, there are few reports on the management and follow-up of patients with the Δ4-3-oxosteroid 5β-reductase (AKR1D1) deficiency. The retrospective analysis of the responses to oral bile acid replacement therapy with chenodeoxycholic acid (CDCA) in patients with this bile acid synthesis disorder we hypothesized will increase our understanding of the disease progression and permit evaluation of this treatment regimen as an alternative to the Food and Drug Administration (FDA) approved drug cholic acid

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