

Towards an optimization of the management of endocrine complications of thalassemia.

El Kholly M, Elsedfy H, Soliman A, Anastasi S, Raiola G, De Sanctis V.

Abstract

Abstract Children with thalassemia are living longer due to better care. Physicians dealing with this group of patients now have to contend with new challenges resulting from iron overload. Endocrine complications represent the most common morbidities encountered. To provide a better quality of life, these complications have to be addressed in a consistent way. For this purpose, we have compiled a set of recommendations to help physicians provide the best care possible to these patients.

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Review Article

Continuous glucose monitoring system and new era of early diagnosis of diabetes in high risk groups

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ABSTRACT

Continuous glucose monitoring (CGM) systems are an emerging technology that allows frequent glucose measurements to monitor glucose trends in real time. Their use as a diagnostic tool is still developing and appears to be promising. Combining intermittent glucose self-monitoring (SGM) and CGM combines the benefits of both. Significant improvement in the treatment modalities that may prevent the progress of prediabetes to diabetes have been achieved recently and dictates screening of high risk patients for early diagnosis and management of glycemic abnormalities. The use of CGMS in the diagnosis of early dysglycemia (prediabetes) especially in high risk patients appears to be an attractive approach. In this review we searched the literature to investigate the value of using CGMS as a diagnostic tool compared to other known tools, namely oral glucose tolerance test (OGTT) and measurement of glycated hemoglobin (HbA1C) in high risk groups. Those categories of patients include adolescents and adults with obesity especially those with family history of type 2 diabetes mellitus, polycystic ovary syndrome (PCO), gestational diabetes, cystic fibrosis, thalassemia major, acute coronary syndrome (ACS), and after renal transplantation. It appears that the ability of the CGMS for frequently monitoring (every 5 min) glucose changes during real-life settings for 3 to 5 days stretches the chance to detect more glycemic abnormalities during basal and postprandial conditions compared to other short-timed methods.

Key words: Continuous glucose monitoring system, Glycated hemoglobin (HbA1C), obesity, oral glucose tolerance test, thalassemia

Is priming with sex steroids useful for defining patients who will benefit from GH treatment?

[De Sanctis V](#), [Soliman AT](#), [Yassin M](#), [Di Maio S](#).

Abstract

Classic criteria for diagnosing GHD include: short stature (height below the third percentile), slow growth velocity, delayed bone age and failure to produce growth hormone in response to two provocative tests. While provocation tests can diagnose complete GHD, debate still exists about of what constitutes a normal or a subnormal GH response in subjects with "idiopathic" short stature or constitutional delay of growth and puberty. It has been suggested that in children with intermediate GH responses to pharmacologic stimuli, a pre-treatment with sex steroids priming may be of value in enhancing the GH response and in helping to clarify the diagnosis, particularly in children with delayed onset of puberty. Nevertheless, the use of priming with sex steroids prior to GH stimulation test in the peripubertal period is still controversial because it is considered an "unphysiologic method" and may mask children with transient GHD. Further studies and uniform guidelines are needed before solving this intriguing puzzle.

Comment in

Editor's commentary: sex hormone priming. [Pediatr Endocrinol Rev. 2014]



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Original article

Longitudinal Study on Liver Functions in Patients with Thalassemia Major before and after Deferasirox (DFX) Therapy

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Abstract. By performing regular blood transfusion and iron chelation therapy, most patients with beta thalassemia major (BTM) now survive beyond the third decade of life. Liver disease is becoming an important cause of morbidity and mortality in these patients. Chronic hepatitis and/or severe iron overload are both important causes of liver pathology. Iron chelation with desferrioxamine (DFO) reduces excessive body iron, but its efficacy is limited by poor compliance and dose related toxicity. The recent use of Deferasirox (DFX), an oral single dose therapy, has improved the compliance to chelation.

Aims: To study the long-term liver functions in BMT patients, seronegative for liver infections before versus after DFX treatment in relation to ferritin level.

Methods: Only BTM patients with hepatitis negative screening (checked every year) and on treatment with DFO for at least five years and with DFX for four years were enrolled. Liver function tests including serum bilirubin, alanine transferase (ALT), aspartate transferase (AST), albumin, insulin-like growth factor – I (IGF-I) and serum ferritin concentrations were followed every six months in 40 patients with BTM.

Results: DFX treatment (20 mg/kg/day) significantly decreased serum ferritin level in patients with BTM; this was associated with a significant decrease in serum ALT, AST, ALP and increase in IGF-I concentrations. Albumin concentrations did not change after DFX treatment. ALT and AST levels were correlated significantly with serum ferritin concentrations ($r = 0.45$ and 0.33 respectively, $p < 0.05$). IGF-I concentrations were correlated significantly with serum ALT ($r = 0.26$, $p = 0.05$) but not with AST, ALP, bilirubin or albumin levels.

The negative correlation between serum ferritin concentrations and ALT suggests that the

