PAEDIATRIC

PE-01

GRAVES DISEASE IN CHILDREN AND ADOLESCENTS: PROGRESSION FROM HYPERTHYROIDISM TO SPONTANEOUS HYPOTHYROIDISM

https://doi.org/10.15605/jafes.036.S94

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INTRODUCTION
Graves' disease (GD) is an autoimmune disorder characterized by hyperthyroidism caused by the presence of thyroid stimulating-antibody. In adult patients with GD, approximately 5-20% of patients eventually progress to hypothyroidism after a period of remission of more than 10 years. Possible mechanisms for the development of spontaneous hypothyroidism are the development of TSH-blocking antibodies or a chronic autoimmune process similar to Hashimoto thyroiditis. In children, whether a subset of patients' progress to hypothyroidism is unclear.

RESULTS
We present three cases of pediatrics GD who progress from hyperthyroidism to hypothyroidism.

Case 1: 17-year-old girl. Treated with carbimazole for 6 years. Defaulted follow up for 1 year, self-prescribed carbimazole. Severely hypothyroidism when re-presented (FT4 3.7 pmol/, TSH 179.9 mIU/L). Currently on L-thyroxine 100 mcg daily past 1 year. At presentation TRAb 28.1 U/L (<1), Anti-TG >4000 U/ml (<1), Anti-TPO >929 U/ml (<1). At remission TSI 120 IU/L (<0.55), anti-TG 1313 U/ml(<1), anti-TPO >972 U/ml(<1)

Case 2: 11-year-old girl. Treated with carbimazole 3 years. Developed subclinical hypothyroidism (FT4 8.4 pmol/L and TSH 7.455 mIU/mL) after 1 year of remission. Spontaneous normalization of TFT after 2 months currently euthyroid. At presentation TRAb 20 U/L (<1), Anti-TG 1425 U/ml, Anti-TPO >986 U/ml.At remission:TRAb 0.46IU/L(<1)

Case 3: 10-year-old girl presented at age of 8 years with weight loss over 4 months. Treated with carbimazole for 1 year. Developed hypothyroidism (FT4 8.9 pmol/L, TSH 13.45 mIU/mL) after 8 months of remission. Currently on L-thyroxine 25 mcg for duration of 3 months. At presentation TRAb 151 IU/L (N<0.55), Anti-TG 408.9U/ml (n<1), Anti-TPO>988 U/ml (N <1). At remission,TRAb 24.2 IU/L(<1),Anti-TG:507 U/ml,Anti-TPO: >986 U/ml.

CONCLUSION
Our cases demonstrate that there are a proportion of children with GD in remission that develop spontaneous hypothyroidism. Highly elevated levels of anti-TPO could be a predictive risk factor for this.

PE-02

RECOMBINANT IGF-1 USE IN SIBLINGS WITH LARON SYNDROME: FIRST 2 CASES TREATED IN MALAYSIA

https://doi.org/10.15605/jafes.036.S95

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INTRODUCTION
Laron Syndrome (LS) is a rare cause of extreme poor growth in children due to the mutation of GHR gene. It is characterised by postnatal growth failure, with midface hypoplasia and obesity. Growth hormone level is normal or elevated with low IGF 1 value. Severe short stature is the major disability in untreated adults. Recombinant IGF 1 hormone (rIGF1) is the only approved treatment since 2007.

RESULTS
We present the case of 2 siblings, whom parents are consanguineous. H presented at 11.5 years old with short stature and being obese; height 121 cm (-3.39 SDS), weight 41.8kg (+0.8 SDS) and BMI 28.55kg/m2 (+2.91 SDS). His birth weight and length were 3kg and 50 cm (-0.5 SDS and +0.3 SDS). Genetic test confirmed homozygous mutation at the GHR gene and presence of heterozygous gene mutation in both parents. The youngest sibling, K was born with birth weight of 3.2kg (+0.98 SDS). At 2.9 years old, K was severely stunted. His height and weight were 64.6cm (-7.67 SDS) and 6.18kg (-8.95 SDS). Both patients scored 4 out of 5 on Savage Scoring System. Recombinant IGF1 (mecasermin) was initiated at the age of 12.7 years and 4.1 years, respectively. At 10 months post rGH1, all growth parameters improved remarkably. Pre- and post-treatment height velocity and serum IGF 1 for H and K