PAEDIATRIC

PE-01

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

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

Farizan G,[1] Jeanne SL Wong,[2] Nalini M Selveindran,[2] Janet YH Hong[2]
[1]*Department of Paediatrics, Hospital Putrajaya, Malaysia*
[2]*Paediatric Endocrine Unit, Hospital Putrajaya, Malaysia*

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

Nurshadia Samingan,[1,2] Meenal Mavinkurve,[3] Mazidah Noordin,[4] Annie Leong,[5] Azriyanti Anuar Zaini,[1,2] Siti Zarina Yaakop,[1] Muhammad Yazid Jalaludin[1,2]
[1]*Department of Paediatrics, University Malaya Medical Centre, Kuala Lumpur, Malaysia*
[2]*Department of Paediatrics, University Malaya, Kuala Lumpur, Malaysia*
[3]*Department of Paediatrics, International Medical University, Seremban, Malaysia*
[4]*Department of Paediatrics, University Teknologi Mara (UiTM), Shah Alam, Malaysia*
[5]*Department of Paediatrics, Hospital Putrajaya, Malaysia*

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

were (4.2cm vs 14.8cm)/year and (4cm vs 17.6cm)/year, (47 vs 329) Âμmol/L and (<15 vs 205) Âμmol/L, respectively. Hypoglycaemia, a common side effect of treatment was not reported. H's percentage body fat and muscle mass improved from 54.1% to 52% and 10.8kg to 12.4kg. One year treatment for both siblings' costs RM751,500 (USD 182,624.57).

CONCLUSION

LS is rare, yet a treatable cause for severe short stature. Albeit the exorbitant cost, treatment offers positive outlook.

PE-03

VITAMIN D DEFICIENCY RICKETS – A CASE SERIES: 'A TIP OF THE ICEBERG'

https://doi.org/10.15605/jafes.036.S96

Naveen N,[1] Cheah YK,[1] Jeanne WSL[2]
[1]*Department of Paediatrics, Hospital Tuanku Ja'afar Seremban, Malaysia,*
[2]*Department of Paediatrics, Hospital Putrajaya, Malaysia*

INTRODUCTION

Vitamin D deficiency is the most common cause of rickets worldwide. In Malaysia, owing to the abundant sunlight exposure, it is believed to be uncommon, however it is likely to be under-reported. In addition, dietary calcium deficiency is an important cause of nutritional rickets in children above 1 year old in developing countries.

RESULTS

We report a case-series of 4 unrelated Malaysian children (aged between 1 to 3) born in Istanbul, Turkey presented with the classical clinical features and biochemical changes of rickets. They were all exclusively breast-fed during infancy with poor dietary calcium intake. Their workup showed normal Calcium, high Alkaline Phosphatase, low Vitamin D and high Parathyroid hormone levels, with radiographic changes of fraying and spraying of the wrist, consistent with Vitamin D Deficiency Rickets. Bowing of legs and widening of wrists joints also seen. Low maternal Vitamin D levels also support the diagnosis. All four children were treated with cholecalciferol (vitamin D3) and short-term calcium supplements. The children showed improvements in growth and normalization of biochemical parameters on follow-ups.

CONCLUSION

Meta-analysis in Turkish populations have shown high prevalence of Vitamin D deficiency leading to their national policy of vitamin D supplementation for infants. Our patients in this case series were neither immunized nor received the appropriate supplements during their stay in Istanbul, Turkey. Maternal vitamin D deficiency, restricted sunlight exposure due to clothing style and seasonal variations, poor dietary calcium intake were all the contributing factors to the nutritional rickets in our patients. Maternal Vitamin D levels could serve as an early indicator of possible deficiency if detected early. Awareness amongst our population was scarce, leading to a delay in seeking treatment/intervention. This case series aims to highlight the importance of vitamin D supplementation as well as ensuring adequate dietary calcium in prevention of nutritional rickets.

PE-04

A CLINICAL PROFILE OF MALAYSIAN PRE-SCHOOL CHILDREN WITH TYPE 1 DIABETES: OBSERVATIONS FROM A SINGLE CENTRE

https://doi.org/10.15605/jafes.036.S97

Meenal Mavinkurve,[1,2] Muhammad Yazid Jalaludin,[2,3] Annie Leong,[2] Mazidah Noordin,[2,4] Nurshadia Samingan,[2] Azriyanti Anuar Zaini[2,3]
[1]*Department of Paediatrics, School of Medicine, International Medical University, Seremban, Malaysia*
[2]*Department of Paediatrics, University Malaya Medical Centre, Kuala Lumpur, Malaysia*
[3]*Department of Paediatrics, Faculty of Medicine, University Malaya, Kuala Lumpur, Malaysia*
[4]*Department of Paediatrics, University Teknologi Mara (UiTM), Sungai Buloh, Malaysia*

INTRODUCTION

Type 1 diabetes (T1DM) is the most common form of childhood diabetes in Malaysian children, the median age being 7.6 years. Worldwide, younger children are increasingly being diagnosed with T1DM, especially in the under 5's age group. Vague clinical symptoms may lead to a protracted presentation and increase the risk of severe complications. This study aims to report the clinical characteristics of Malaysian pre-school (<7 years) children with T1DM .

METHODOLOGY

A retrospective review of the demographic and clinical data on children < 7 years of age diagnosed with T1DM at the University of Malaya Medical Centre between January 1st 2010-Dec 31st 2019 was conducted.