

RESULT

Data was available for 10 patients. Most of them had malignancy related SIADH. Based on our protocol, all patients were given an initial dose of Tolvaptan 7.5 mg (compared to manufacture recommendation of 15 mg) to avoid the risk of Na overcorrection. The mean baseline Na was 118 mmol/L (range 110-123 mmol/L). The mean rise in Na at 24 hours was 7 mmol/L (range 1-11 mmol/L). 9 patients had Na increment of at least 5 mmol/L at 24 hours. The remaining 1 patient had increment of Na level of only more than 1 mmol/L at 24 hours but responded to Tolvaptan 15 mg subsequently. None of the patients had overcorrection of Na (more than 12 mmol/day over 24 hours) or development of osmotic demyelination syndrome.

CONCLUSION

This study demonstrated that inpatient tolvaptan use resulted in favourable outcomes. Our local Tolvaptan protocol is safe and effective in the treatment of SIADH-related hyponatraemia.

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ENDOCRINOPATHIES IN ADULT PATIENTS WITH TRANSFUSION-DEPENDENT THALASSEMIA IN HOSPITAL SULTANAH BAHYAH

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LW Wong, Nor Shaffinaz Yusoff Azmi Merican, Noor Rafhati Adyani Abdullah, Norasmidar Aziz, Shartiyah Ismail

Hospital Sultanah Bahiyah, Malaysia

INTRODUCTION

Thalassaemia is the most common hereditary haematological disorder in Malaysia. Patients with transfusion-dependent thalassaemia (TDT) treated with frequent blood transfusion are typically at risk for developing multiple complications like endocrinopathies. Our primary objective is to determine the screening rate of endocrinopathies among our TDT patients. We also aimed to determine the prevalence of endocrinopathies and factors correlated with endocrinopathies.

METHODOLOGY

This was a retrospective evaluation of all 113 patients with TDT who were under the care of the Haematology clinic, Hospital Sultanah Bahiyah with follow-up 6 months prior to data collection. Relevant data were retrieved from electronic medical records. Data collection was done in April 2023. Statistical analyses were performed using SPSS version 11.0.

RESULT

Median age of our cohort is 31 years (Interquartile range 18 years), and 50.4% of the patients were female. Majority of the patients (92%) received at least one endocrinopathy screening. The most commonly screened endocrinopathies were hypothyroidism (82%), followed by diabetes mellitus (66%) and hypoparathyroidism (53%). About 39% of our patients have at least one endocrine complication. Of these, 24% had one endocrinopathy, 11.5% had two endocrinopathies while 3.6% had 3 or more endocrinopathies. Growth failure (58%) was the most prevalent endocrinopathy among our cohort, followed by hypocortisolism (50%) and osteopenia (48%) while prevalence of hypothyroidism was 15%. There were significant correlations between ferritin level and liver iron concentration (LIC) [$p < 0.01$] and cardiac iron concentration (CIC) [$p < 0.01$]. No significant correlation was found between ferritin level and the presence of endocrinopathies, or between LIC and CIC with the presence of endocrinopathies.

CONCLUSION

There was a low screening rate for endocrinopathies among our patients, particularly for osteoporosis, growth failure, hypocortisolism and hypogonadism. This audit shall serve to raise the awareness of healthcare practitioners to screen for endocrinopathies among patients with TDT.

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SUCCESSFUL PREGNANCY IN A HOMOZYGOUS FAMILIAL HYPERCHOLESTEROLEMIA PATIENT REQUIRING LIPID APHERESIS AND MULTIDISCIPLINARY TEAM MANAGEMENT

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Siau Chenn Khong, Ashcwiniswarie Gunasegaran, Nurul Atikah Hamza, Hazwani Aziz, Sadanah Aqashiah Mazlan, Elliyyin Katiman

Hospital Kajang, Malaysia

INTRODUCTION/BACKGROUND

Familial hypercholesterolemia (FH) is an inherited disorder of lipid metabolism mainly due to the mutation of the low-density lipoprotein (LDL)-receptor gene (LDLR). The more severe homozygous form of the disease is characterised by premature atherosclerotic disease before 20 years old. Pregnancy in women with homozygous familial hypercholesterolemia (HoFH) can be fatal to both mother and foetus. Since Lipid Apheresis (LA) is scarce in Asia and statin generally is contraindicated in pregnancy, treatment to keep low-density lipoprotein-cholesterol (LDL-C) at low levels remain challenging during pregnancy, necessitating a multidisciplinary team approach. We illustrate the case