

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.

EP_A032

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

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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.

EP_A033

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

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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

of a 24-year-old primigravida with HoFH who underwent LA and successfully delivered her baby at 35 weeks age of gestation.

CASE

A 24-year-old female diagnosed with HoFH at 7 years old, presented at 7 weeks pregnancy with hyperemesis gravidarum. She previously required regular plasma exchange from the age of 8 but subsequently defaulted treatment at 16 years old. Clinical examination revealed widespread multiple xanthomata over both hands, feet and elbows. Her baseline total cholesterol was 15 mmol/L and LDL-C was 13.2 mmol/L. She was initiated on bi-weekly plasma exchange. However, she developed intradialytic hypotension complicated by fistula failure following a second exchange, which necessitate double filtration plasmapheresis (DFPP), which is more specific for lipid apheresis. The LDL-C levels were reduced by an average of 46% following each treatment. Her pregnancy was complicated by two hospitalisations for suspected Acute Coronary Syndrome. Cardiology referral was made for re-assessment of coronary arteries. Fortunately, echocardiography and dobutamine stress test both showed normal findings. At 35 weeks of gestation, the patient successfully delivered a healthy baby boy weighing 1.6 kg via emergency caesarean section for foetal complication with good Apgar score.

CONCLUSION

This case demonstrated a favourable pregnancy outcome when LA along with good multidisciplinary support was utilized in a pregnant patient with HoFH.

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THE USE OF THERAPEUTIC PLASMA EXCHANGE IN A PATIENT WITH RECURRENT SEVERE HYPERTRIGLYCERIDEMIA-INDUCED ACUTE PANCREATITIS

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INTRODUCTION/BACKGROUND

Severe hypertriglyceridemia (HTG)-induced acute pancreatitis is defined by clinical, laboratory and radiographic evidence of acute pancreatitis with triglyceride (TG) levels of >11.2 mmol/L in the absence of other causative factors. We report a case of therapeutic plasma exchange (TPE) used in recurrent severe HTG-induced acute pancreatitis who failed conventional treatment.

CASE

The patient is a 28-year-old female with poorly controlled Type 2 diabetes mellitus diagnosed 4 years ago with HbA1c range of 10-13% despite on Insulin Actrapid 30 u three times daily, Insulin glargine 36 u daily and Metformin 1g twice daily. Her TG levels remained elevated despite being on daily 145 mg of Fenofibrate, 40 mg of Rosuvastatin, and 4 gm of Omega-3-free fatty acid. Xanthelasma, tuberous or tendon xanthomata were absent. She denied a family history of hypertriglyceridemia. She was non-alcoholic and her thyroid screen was normal. Obesity and poor compliance with lifestyle changes and medications alongside poorly controlled diabetes contributed to severe hypertriglyceridemia. She had recurrent admissions for severe HTG-induced acute pancreatitis within the past 2 years. During each admission, she was given supportive treatment including fasting with bowel rest, analgesia, intravenous hydration, and insulin infusion. She failed to respond to conservative measures and required TPE for 3 of her 6 admissions. Her TG level was >64 mmol/L during these 3 admissions with persistent severe abdominal pain lasting more than 48-72 hours despite fentanyl infusion. Fresh frozen plasma was used as replacement fluid during each TPE session. TG levels dropped by 80-85% after a single TPE with TG levels on discharge decreased to a range of 2.3-5.5 mmol/L.

CONCLUSION

This case highlights the potential utility of TPE during acute pancreatitis by rapidly decreasing TG levels and reducing inflammatory cytokines. However, the TPE effect is transient and the patient requires adequate lipid-lowering treatment to achieve lasting effects.

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HEPATOMA-ASSOCIATED NON-ISLET CELL TUMOR HYPOGLYCEMIA: A CASE REPORT

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INTRODUCTION/BACKGROUND

Non-islet cell tumour hypoglycaemia (NICTH) is a rare condition due to excessive secretion of insulin-like growth factor-2 (IGF-2) or pro-IGF-2. NICTH is commonly associated with hepatocellular carcinomas.

CASE

We reviewed case notes, investigation results, imaging studies and discussed treatment options based on literature review.