OP_P005

OUTCOME OF CONGENITAL HYPERINSULINISM (CHI): A SINGLE-CENTRE EXPERIENCE

https://doi.org/10.15605/jafes.038.S2.124

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INTRODUCTION

Congenital hyperinsulinism (CHI) is a condition that results in recurrent hypoglycaemia due to inappropriate insulin secretion. This condition is generally rare; however, it is still a common cause of persistent hypoglycaemia beyond infancy.

METHODOLOGY

This is a cross-sectional study of the patients attending the paediatric endocrinology unit in the National Centre for Child Health & Development (NCCHD), Tokyo, Japan from March 2022 to March 2023. Our study aimed to analyse the characteristics of the CHI patients who are in remission as compared to those who still have persistent disease.

RESULT

There was a total of 39 patients, with 23(59%) patients who were male. The mean age was 10.9 (2.3 – 31.75) years old and the majority of the patients were of Japanese ethnicity, 37 (94.9%). At the time of study, 28 (71.8%) patients still had persistent CHI, while the remaining were in remission.

There was a total of 10 patients who had surgery in our cohort. Among those patients who had surgery, 8 (80%) are already in remission. The majority of the CHI patients, 29 (74%), were managed with medical therapy, and among these, only 3 patients (10.3%) are in remission. The patients who still have persistent disease are still dependent on nutritional and/or medical therapy. Among the patients who still have persistent disease, 23 patients (82.1%) require monotherapy while the remaining 5 patients (17.9%) require 2 or more therapy to maintain the blood glucose within control. Long-term complications were observed within our cohort including diabetes and dysregulated glycemia, pancreatic insufficiency and neurological effects.

CONCLUSION

In conclusion, CHI is a challenging disease to manage, not only for the paediatric endocrinologists, but most importantly, for the patients and their families. This condition not only requires multidisciplinary management, but also close follow-up for long-term complications.