Glucose variability may be unrecognised in CHS, particularly in children with features of Autonomic nervous system dysfunction (ANS). This report highlights the occurrence of hyperglycaemia as well as hypoglycaemia in CHS. Given the challenges of recognising hypoglycaemia based on clinical symptomatology the use of CGM may be an appropriate method of screening. The observed normoglycaemia during fasting with increased post-prandial BGL variability is consistent with a dynamic dysregulation in the central autonomic control of insulin secretion. ANSD is likely to be influencing the responses that co-ordinate glucose delivery across the gut and peripheral insulin mediated glucose disposal. Dietary modifications may be more effective than Diazoxide in managing hypoglycaemia. The long-term consequences of dysregulated glucose homeostasis in this group are unknown.