

Heterogeneous Presentation of Paediatric Hyperglycaemic Hyperosmolar State – A Case Series



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INTRODUCTION

Hyperosmolar hyperglycaemic state (HHS) is a rare, but recognised, life-threatening clinical entity in children with type 2 diabetes (T2DM).

- Diagnostic criteria for HHS include: Blood glucose >33.3mmol/L, Serum osmolality >320mOsm/L, absence of significant ketosis and acidosis (serum bicarbonate >15mmol/L).
- HHS is also reported as presenting feature in other types of diabetes and metabolic disorders. The estimated mortality in HHS is 10-20%, 10 times higher than DKA.
- The mainstay of management involves aggressive fluid therapy with insulin and managing complications namely rhabdomyolysis, multi-organ failure, cerebral oedema and malignant hyperthermia.

OBJECTIVE

To report three cases, where HHS was the presenting feature, only one of which had confirmed T2DM, highlighting the challenges for early diagnosis and management.

Case 1

A 16-year-old-female of Asian ancestry, with background of undiagnosed neurodegenerative disease, immobility, pubertal delay, hyperphagia and obesity (BMI 42) presented to the emergency department with a decreased level of consciousness following a history of tonsillitis (GCS 9/15). On examination she had marked acanthosis nigricans. Investigation revealed mild acidosis pH-7.27 with blood glucose (BG) of 33mmol/L, initially managed as DKA. She progressively deteriorated with GCS 6/15, polyuria and hypernatraemic dehydration. Subsequent investigations revealed, serum-osmolality (392mOsm/kg), with no urinary ketones confirming HHS and T2DM. She had a complicated course with acute kidney injury, vascular thrombosis, cerebral oedema, prolonged recovery phase and loss of orality requiring supportive tube feeding.

Case 2

Previously fit and well, 4.5 year old male child of Black-African origin with BMI 16.5kg/m², presented with acute collapse (GCS 3/15), severe metabolic acidosis (pH 6.8, Lactate 13.7mmol/L) and abdominal distension. Laboratory investigation revealed; hyperglycaemia, absent ketones, hypernatraemia and raised serum-osmolality(400mOsm/kg). He stopped fluid and insulin therapy after 4 days of initiation, had an uncomplicated complete recovery with return to normal mental state in 9 days. The aetiology of presentation was unclear but had an overlay of features of DKA and HHS at presentation*.

Case 3

An 8-year-old Caucasian girl with background of anaemia and global developmental delay with quadriplegic cerebral palsy, Haemophagocytic Lymphohistiocytosis, receiving pulsed steroid therapy, presented with drowsiness and severe dehydration. Management involved aggressive isotonic fluid resuscitation and insulin therapy. She had an uncomplicated recovery to premorbid state in 6 weeks time. There was no clinical or biochemical evidence of T2DM on follow up at 3 months time, from the initial presentation.

Summary of investigations

Investigations	Case 1	Case 2	Case 3
pH	7.27	6.8	7.32
Blood glucose (mmol/L)	33	47	38
Sodium(mmol/L)	172	163	152
Serum-Osmolality(Osm/Kg)	392	400	368
HbA1C(mmol/mol)	104	<40	82
Blood Ketones(mmol/L)	1.8	Negative	2.4

CONCLUSIONS

1. This heterogeneous case series includes a new-onset T2DM, a steroid induced HHS and transient HHS with complete recovery. The latter two have not been previously described in the literature.
2. Significant neurodevelopmental delay in two of our patients delayed diagnosis similar to other cases described in the literature.
3. There is imperative need for increased awareness and evidence-based paediatric guidelines for management of HHS to avert adverse outcomes.

References

*Zeitler P, Haqq A, Rosenbloom A, Glaser N; Drugs and Therapeutics Committee of the Lawson Wilkins Pediatric Endocrine Society. Hyperglycemic hyperosmolar syndrome in children: pathophysiological considerations and suggested guidelines for treatment. *J Pediatr* 2011;158:9-14