British Inherited Metabolic Disease Group

RECURRENT HYPOGLYCAEMIA

Background

Hypoglycaemia is a feature of many conditions, including well defined inherited metabolic diseases and endocrine disorders. It can also occur, for reasons that are not completely understood, in children who appear otherwise healthy, a pattern of biochemical response sometimes referred to as 'ketotic hypoglycaemia' (sometimes also called 'accelerated starvation'). Correct management requires as precise a definition as possible. An accurate diagnosis is most easily made by taking the correct specimens at the time of hypoglycaemia.

Ideally, the following investigations should be done (sometimes known as the 'hyposcreen') at the time of hypoglycaemia. To avoid delays please add clear instructions for the tubes and the quantities of blood required by your local laboratory.

		Tube	Volume
Blood/ Plasma	glucose (in the laboratory)		
	lactate	-	-
	**free fatty acids (= NEFA, non esterified fatty acids)	-	-
	** § 3-hydroxybutyrate	-	-
	insulin	-	-
	cortisol	-	-
	growth hormone		
	quantitative amino acids		
Blood spot	acyl carnitine profile (also in plasma if possible)	blood spot	2 spots*
Urine	ketones and organic acids (collect the next specimen passed)	-	-

NOTES

* minimum

** If these are not measured by your local laboratory, please collect 2 ml heparinised blood, separate quickly and store the plasma deep frozen so that it can be assayed later. Contact your local specialist centre for advice.

§ A bedside ketone meter may be used as alternative if available

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ADDITIONAL IMPORTANT NOTES

- **The most important blood samples are those for glucose and insulin**, followed by those for cortisol and lactate. If there are difficulties ordering the tests, collect blood into a heparinised tube, separate quickly and store the plasma deep frozen.

- **DO NOT delay correcting the hypoglycaemia if blood is difficult to obtain**. Give glucose 10% 2ml/kg (200 mg/kg) followed by a continuous infusion of 4-6mg/kg/min to maintain normoglycaemia

The following may be needed in special circumstances. Discuss with specialist.
Blood/plasma: Pyruvate, Acetoacetate
Ammonia
C-peptide
Glucagon
Growth hormone IGF1, IGF2, IGFBP1 & IGFBP3
Catecholamines

If blood was not obtained acutely, blood spots for acylcarnitine analysis can still be sent when the patient is well.

MANGEMENT
Discuss the further investigations and management with a metabolic specialist or an endocrinologist.

Reference: Ghosh A, Banerjee I, Morris AA. Recognition, assessment and management of hypoglycaemia in childhood. Arch Dis Child 2016;101:575–80.

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