



Metabolic Storage Disorders of the Nervous System

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Case-Based Questions (please see page 3 for answers)

1.	Which of the following statements about cell constituent recycling is correct?
a.	Most normal proteins are stable for years.
b.	Glycogen in astrocytes is replaced every 3-5 days.
c.	Myelin lipids are stable for only a few days.

2.	The Luxol fast blue reagent stains which of the following cell constituents?
a.	Myelin basic protein in myelin.
b.	The abnormal storage material in Lafora disease.
c.	The abnormal neuronal storage material in multiple sulfatase deficiency and many of the mucopolysaccharidoses.

3.	Which statement about the mucopolysaccharidoses (MPS) is correct?
a.	MPS are associated with perivascular macrophages that contain lamellar inclusions that resemble "crinkled paper".
b.	In addition to neurological disease, MPS are often associated with coarse facial features and hepato-splenomegaly.
c.	Electron microscopic studies of MPS usually shows crystalline inclusion material in the cytoplasm.

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Correct Answers and Rationales

Question 1 Correct Answer and Rationale: **B. Glycogen in astrocytes is replaced every 3-5 days**

Rationale: Glycogen turnover has been well studied in vivo using radiolabeled glucose infusions. Most proteins are stable for a few days to few months. Most membrane lipids are stable for weeks to months.

Question 2 Correct Answer and Rationale: **C. The abnormal neuronal storage material in multiple sulfatase deficiency and many of the mucopolysaccharidoses.**

Rationale: LFB stains gangliosides in myelin. Lafora disease is associated with accumulation of polyglucosans, which stain with periodic acid Schiff method.

Question 3 Correct Answer and Rationale: **B. In addition to neurological disease, MPS are often associated with coarse facial features and hepato-splenomegaly.**

Rationale: A is a feature of Gaucher disease. The inclusions in MPS are typically fibrillogranular or lamellar within lysosomes.