Lippincott Biochemistry test bank for lectures (1-10)

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وَتَوَكَّلْ عَلَى الْحَيِّ الَّذِي لَا يَمُوتُ وَسَبِّحْ بِحَمْدِهِ ۚ وَكَفَىٰ بِهِ
بِذُنُوبِ عِبَادِهِ خَبِيرًا

Pentose phosphate pathway (PPP)

In preparation for a trip to an area of India, a young male is given an antimalarial drug prophylactically. Several days after initiation of this therapy he develops jaundice and is diagnosed with anemia. A low level of which of the following is a consequence of the most likely enzyme deficiency and the underlying cause of the patient's presentation?

A. Glucose 6-phosphate
B. Oxidized form of nicotinamide adenine dinucleotide
C. Reduced form of glutathione
D. Ribose 5-phosphate

Correct answer = C. Glutathione (G-SH) is essential for red cel integrity and is maintained in this reduced (functional) form by nicotinamide adenine dinucleotide phosphate (NADPH)-dependent glutathione reductase. The NADPH is from the oxidative portion of the pentose phosphate pathway. Individuals with a deficiency of the regulated enzyme of this pathway, glucose 6-phosphate dehydrogenase (G6PD), have a decreased ability to generate NADPH and, therefore, a decreased ability to keep G-SH reduced. When treated with some antimalarials that induce an oxidant stress, some patients with G6PD deficiency develop a hemolytic anemia. Levels of glucose 6-phosphate are not altered. Nicotinamide adenine dinucleotide (NAD[H]) is neither produced by the pathway nor used as a coenzyme by G-SH reductase. A decrease in ribose 5-phosphate does not cause hemolysis.

Low blood pressure (hypotension), is a sign of septic shock, resulting from a severe inflammatory response to a bacterial infection. Based on this information, a likely cause of this hypotension is:

A. Activation of endothelial nitric oxide synthase causing a decrease in nitric oxide.
B. The long half-life of nitric oxide promotes long-term, excess vasoconstriction.
C. Lysine, the nitrogen source for nitric oxide synthesis, is deaminated by bacteria.
D. Bacterial endotoxin promoting iNOS synthesis causing increased NO production.

Correct answer = D. Overproduction of short-lived nitric oxide (NO) by calcium independent, inducible nitric oxide synthase (iNOS) results in excessive vasodilation, leading to hypotension. The endothelial enzyme (eNOS) is constitutive and produces low levels of NO at a consistent rate. NOS uses arginine, not lysine, as the source of the nitrogen

In males who are hemizygous for glucose 6-phosphate dehydrogenase deficiency, pathophysiologic consequences are more apparent in red blood cels than in other cels such as in the liver. The best explanation for these findings is that:

A. Excess glucose 6-phosphate in the liver, but not in red blood cels, can be channeled to glycogen, thereby averting cellular damage.

B. Liver cels, in contrast to red blood cels, have alternative mechanisms for supplying the reduced nicotinamide adenine dinucleotide phosphate required for maintaining cel integrity.

C. Red blood cel production of ATP required to maintain cel integrity depends exclusively on the shunting of glucose 6-phosphate to the pentose phosphate pathway.

D. In contrast to liver cels, red cel glucose 6-phosphatase activity decreases the level of glucose 6-phosphate, resulting in cel damage.

Correct answer = B. Cellular damage is directly related to decreased ability of the cel to regenerate reduced glutathione, for which large amounts of reduced nicotinamide adenine dinucleotide phosphate (NADPH) are needed, and red blood cels have no means other than the pentose phosphate pathway of generating NADPH. It is decreased product (NADPH), not increased substrate (glucose 6-phosphate), that is the problem. Red blood cels do not have glucose 6-phosphatase. The pentose phosphate pathway does not generate ATP.

Lipid metabolism

Which one of the following statements about lipid absorption from the intestine is correct?

A. Dietary triacylglycerol must be completely hydrolyzed to free fatty acids and glycerol before absorption.

B. The triacylglycerol carried by chylomicrons is degraded by lipoprotein lipase, producing fatty acids that are taken up by muscle and adipose tissues and glycerol that is taken up by the liver.

C. Fatty acids that contain ≤12 carbon atoms are absorbed and enter the circulation primarily via the lymphatic system.

D. Deficiencies in the ability to absorb fat result in excessive amounts of chylomicrons
in the blood

Correct answer = B. The triacylglycerols (TAG) in chylomicrons are degraded to fatty acids (FA) and glycerol by lipoprotein lipase on capilary endothelial surfaces in muscle and adipose tissue, thus providing a source of FA to these tissues for degradation or storage and providing glycerol for hepatic metabolism. In the duodenum, TAG are degraded to one 2-monoacylglycerol + two free FA that get absorbed. Medium- and short chain FA enter directly into blood (not lymph), and they neither require miceles nor get packaged into chylomicrons. Because chylomicrons contain dietary lipids that were digested and absorbed, a defect in fat absorption would result in decreased production of chylomicrons

A 2-year-old female is brought to the physician because of recurrent respiratory tract infections, weight loss, and foul-smelling diarrhea. This patient most likely has a defective secretion in which of the following?

A. Cholecystokinin
B. Pancreatic enzymes
C. Chylomicron
D. Secretin

Correct answer: B. This patient most likely has cystic fibrosis (CF) which causes defective secretion of pancreatic enzymes such as lipase and colipase due to mutations in the cystic fibrosis transmembrane conductance receptor (CFTR). These enzymes are important for digestion and absorption of lipids. Cholecystokinin and secretin are released from enteroendocrine cels. Although they are important for lipid digestion and absorption, they are not defective in CF. Chylomicron formation and release into lymphatic system is not affected in CF

A 45-year-old female is brought to the emergency department due to acute pain, nausea, and vomiting. Computed tomography indicates acute pancreatitis that leads to an increased activation of trypsin. Which of the following is most likely activated in this condition?

A. Gastric lipase
B. Pancreatic lipase
C. Lysophospholipase
D. Colipase

Correct answer: D. Colipase is secreted as the zymogen, procolipase, which is activated in the intestine by trypsin. Colipase is important for pancreatic lipase for hydrolyzing triacylglycerols. Gastric lipase hydrolyze short and medium chain fatty acids in milk, especialy important for infants and patients with pancreatic insufficiency. Lysophospholipase is important for the digestion of phospholipids.

A 22-month-old child is brought to the physician by her parents because of refusal to feed, chronic diarrhea, abdominal distension, and weight loss. She is diagnosed with chylomicron retention disease, which prevents the release chylomicrons into the lymphatics. This patient most likely has a deficiency in which of the following vitamins? *(I'm not sure whether this question is part of the material)*

A. Ascorbic acid
B. Beta-carotene
C. Folate
D. Pyridoxine

Correct answer: B. Chylomicron is important for the absorption of fat-soluble vitamins: vitamin A, D, E, and K. Beta-carotene is a provitamin A that is packaged into chylomicron before its release into the lymphatics. Ascorbic acid is vitamin C, folate is vitamin B9, and pyridoxine is vitamin B6. These three vitamins are water soluble

Fatty Acid, Triacylglycerol, and Ketone Body Metabolism

When oleic acid, 18:1(9), is desaturated at carbon 6 and then elongated, what is the correct representation of the product?

A. 19:2(7,9)
B. 20:2(ω-6)
C. 20:2(6,9)
D. 20:2(8,11)

Correct answer = D. Fatty acids are elongated in the smooth endoplasmic reticulum by adding two carbons at a time to the carboxylate end (carbon 1) of the molecule. This pushes the double bonds at carbon 6 and carbon 9 farther away from carbon 1. The 20:2(8,11) product is an ω-9 (n-9) fatty acid.

A 4-month-old child is being evaluated for fasting hypoglycemia. Laboratory tests at admission reveal low levels of ketone bodies (hypoketonemia), free carnitine, and long chain acylcarnitines in the blood. Free fatty acid levels in the blood were elevated.

Deficiency of which of the following would best explain these findings?

A. Adipose triglyceride lipase
B. Carnitine transporter
C. Carnitine palmitoyltransferase-I
D. Long-chain fatty acid dehydrogenase

Correct answer = B. A defect in the carnitine transporter (primary carnitine deficiency) would result in low levels of carnitine in the blood (as a result of increased urinary loss) and low levels in the tissues. In the liver, this decreases fatty acid oxidation and ketogenesis. Consequently, blood levels of free fatty acids rise. Deficiencies of adipose triglyceride lipase would decrease fatty acid availability. Deficiency of carnitine palmitoyltransferase I would result in elevated blood carnitine. Defects in any of the enzymes of β-oxidation would result in secondary carnitine deficiency, with a rise in acylcarnitines.

A teenager, concerned about his weight, attempts to maintain a fat-free diet for a period of several weeks. If his ability to synthesize various lipids are examined, which of the following is most likely to be most deficient in his ability to synthesize?

A. Cholesterol.
B. Glycolipids.
C. Phospholipids.
D. Prostaglandins.
E. Triacylglycerol.

Correct answer = D. Prostaglandins are synthesized from arachidonic acid. Arachidonic acid is synthesized from linoleic acid, an essential fatty acid obtained by humans from dietary lipids. The teenager would be able to synthesize al other compounds but, presumably, in somewhat decreased amounts.

A 6-month-old male was hospitalized following a seizure. History revealed that for several days prior, his appetite was decreased owing to a stomach virus. At admission, his blood glucose was 24 mg/dl (age-referenced normal is 60 to 100). His urine was negative for ketone bodies and positive for a variety of dicarboxylic acids. Blood carnitine levels (free and acyl bound) were normal. A tentative diagnosis of medium chain fatty acyl coenzyme A dehydrogenase (MCAD) deficiency is made. In patients with MCAD deficiency, which of the following is most likely explanation for the fasting hypoglycemia?

A. Decreased acetyl coenzyme A production.
B. Decreased ability to convert acetyl coenzyme A to glucose.
C. Increased conversion of acetyl coenzyme A to acetoacetate.
D. Increased production of ATP and nicotinamide adenine dinucleotide

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Correct answer = A. Impaired oxidation of fatty acids <12 carbons in length results in decreased production of acetyl-coenzyme A (CoA), which is the alosteric activator of pyruvate carboxylase, a gluconeogenic enzyme; thus, glucose levels fal. Acetyl CoA can never be used for the net synthesis of glucose. Acetoacetate is a ketone body, and with medium-chain fatty acyl CoA dehydrogenase deficiency, ketogenesis is decreased as a result of decreased production of the substrate, acetyl CoA. Impaired fatty acid oxidation means that less ATP and nicotinamide adenine dinucleotide are made, and both are needed for gluconeogenesis.

A 6-week-old female is brought to the emergency room due to hypotonia and failure to thrive. Physical examination shows dysmorphic facial features and hepatomegaly. Laboratory studies show high levels of very long-chain fatty acids and phytanic acid. Which of the following is the most likely diagnosis?

A. X-linked adrenoleukodystrophy
B. Refsum disease
C. Zelweger syndrome
D. Very-long-chain fatty acids (VLCFA) deficiency

Correct answer = C. Zelweger syndrome is caused by an inability to target matrix proteins to the peroxisome. Therefore, al peroxisomal activities are affected because functional peroxisomes are not formed. As a result, laboratory studies show elevation in both VLCA and phytanic acid in serum. Refsum disease caused by a deficiency of peroxisomal PhyH. This results in the accumulation of phytanic acid in the plasma and tissue. In X-linked adrenoleukodystrophy, the defect is an inability to transport VLCFA into the peroxisome, but other peroxisomal functions, such as α-oxidation, are normal.

Phospholipid, Glycosphingolipid metabolism

An infant, born at 28 weeks' gestation, rapidly gave evidence of respiratory distress. Clinical laboratory and imaging results supported the diagnosis of infant respiratory distress syndrome. Which of the following is the most accurate statement about this syndrome?

A. It is unrelated to the baby's premature birth.
B. It is a consequence of too few type II pneumocytes.
C. The lecithin/sphingomyelin ratio in the amniotic fluid is likely to be high (>2).
D. The concentration of dipalmitoylphosphatidylcholine in the amniotic fluid would be expected to be lower than that of a full-term baby.
E. It is an easily treated disorder with low mortality.
F. It is treated by administering surfactant to the mother just before she gives birth.

Correct answer = D. Dipalmitoylphosphatidylcholine (DPPC or, dipalmitoyl lecithin) is the lung surfactant found in mature, healthy lungs. Respiratory distress syndrome (RDS) can occur in lungs that make too little of this compound. If the lecithin/sphingomyelin (L/S) ratio in amniotic fluid is ≥2, a newborn's lungs are considered to be sufficiently mature (premature lungs would be expected to have a ratio

A 10-year-old male was evaluated for burning sensations in his feet and clusters of small, red-purple spots on his skin. Laboratory studies revealed protein in his urine.Enzymatic analysis revealed a deficiency of α-galactosidase, and enzyme replacement therapy was recommended. Which of the following is the most likely working diagnosis?

A. Fabry disease
B. Farber disease
C. Gaucher disease
D. Krabbe disease
E. Niemann-Pick disease

Correct answer = A. Fabry disease, a deficiency of α-galactosidase, is the only X-linked sphingolipidosis. It is characterized by pain in the extremities, a red-purple skin rash (generalized angiokeratomas), and kidney and cardiac complications. Protein in his urine indicates kidney damage. Enzyme replacement therapy is available.

A 5-year-old child is brought to the pediatrician by his mother due to abdominal distention and pain in his leg. The mother states that her son started having difficulty walking and began to fal repeatedly. Physical examination shows developmental delay and hepatosplenomegaly. Fundoscopic examination shows cherry-red spots in the macula. Which of the following histologic finding of the affected tissue is most likely to confirm the diagnosis? *(I'm not sure whether this question is part of the material)*

A. Shel-like inclusion bodies in neuronal cels
B. Crumpled tissue paper appearance of the cytosol
C. Foamy macrophages in the bone marrow
D. Globoid bodies in macrophages

Correct answer = C. Niemann–Pick disease type B is the most likely diagnosis due to the presence of hepatomegaly, neurologic defects leading to fals and cherry-red areas in the macula. The histologic finding is the foamy appearance of macrophages of the reticuloendothelial system because of sphingomyelin accumulation.