

What's all the fuss about fats?

Name _____

Date _____

PART 1.

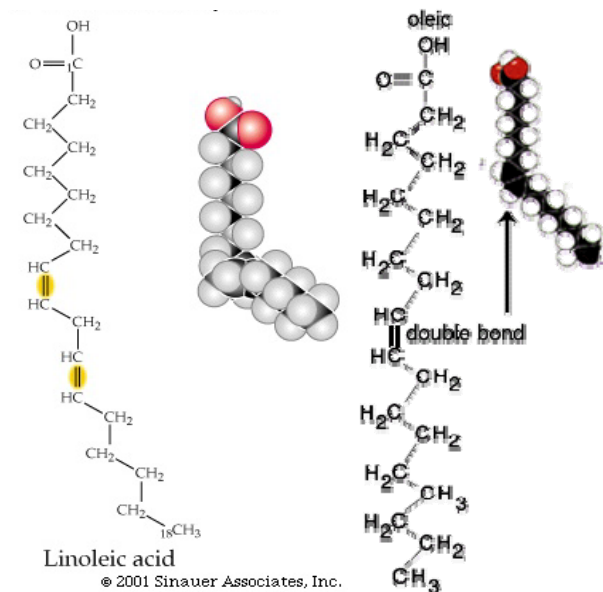
Saturated Fatty Acids vs. Unsaturated Fatty Acids

Saturated fatty acids are saturated with hydrogen atoms which results in **no flexibility** of the chain. These fats are solid at room temperature and are more likely to clog arteries and raise cholesterol levels. These types of fats are found naturally in meat and dairy products.

Unsaturated fatty acids contain double bonds between some carbon atoms reducing the number of hydrogens in the chain. These fats are more flexible and are liquid at room temperature.

They are less stable than saturated fats and easier for our bodies to metabolize. These types of fats are found in oils such as vegetable oil.

Because unsaturated fats are less stable than saturated fats, food manufacturers *hydrogenize* these good fats by adding hydrogen and turning them into the more shelf-life friendly saturated fats, also called trans fatty acids. Trans fats are bad for your body, especially your heart. When looking at nutrition labels, look for foods low in saturated fats and avoid trans fats and hydrogenated or partially hydrogenated fats.

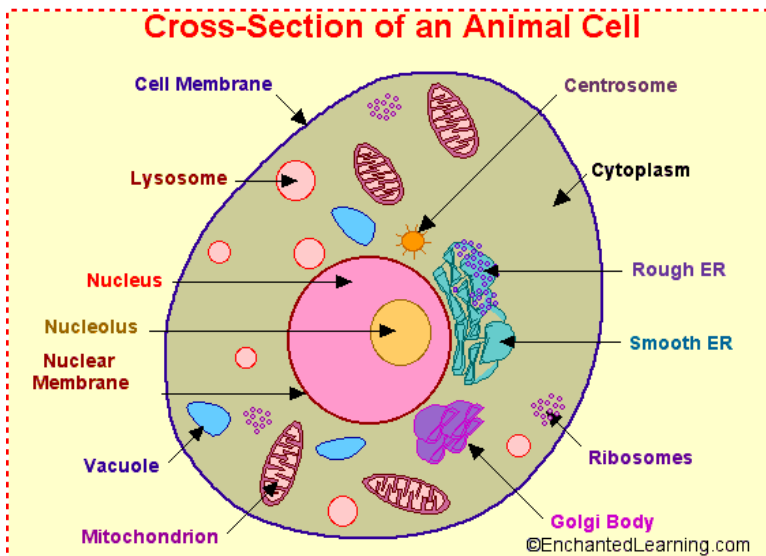


Q1. Using the information you read, explain the difference between saturated and unsaturated fatty acids.

Q2. How could the differences in their structure affect the function of these two fatty acids?

PART 2.

Fats in the cell



The **endoplasmic reticulum (ER)** is an interconnected network of tubules within the cell. The ER has many specialized functions within the cell including the production of long chain fatty acids.

The enzymes in the ER take short chain fatty acids and add on saturated carbon bonds making the chains longer (up to 24 C long). Your body uses these long chain fatty acids

to make hormones, acids in the digestive system, cholesterol in the liver as well as the lipids that make up cell membranes. Although these VLCSFAs are necessary for normal body functions, too much is a bad thing. In a healthy person, when there are too many long chains, they are broken down by special enzymes that only work in the peroxisome.

Q3. What is the function of enzymes in the ER?

Peroxisomes are organelles that resemble lysosomes. Like lysosomes, they have the ability to breakdown larger molecules and cellular waste. These organelles specialize in using a type of protein called an enzyme to break down large molecules.

The enzymes within the peroxisome can't work outside of this organelle and must be delivered from other areas of the cell. These enzymes are transported across the peroxisome membrane using special proteins within the membrane.

The peroxisome is involved in controlling the amount of very long chain saturated fatty acids (VLCSFA) within the cell. **If there are too many of these VLCSFAs, they will be transported to the peroxisome, where the enzymes breakdown these chains 2 C at a time.**

Q4. What is the function of enzymes within the peroxisome?

Q5. How do the ER and peroxisome work together to maintain homeostasis of FA levels within the cell?

ALD patients, like Lornezo Adone, have working ER enzymes and are able to make very long chain saturated fatty acids (VLCSFA) inside their cells. But, **ALD patients lack the necessary membrane proteins needed to transport the enzymes into the peroxisome.** Since the oxidative enzyme is not inside the peroxisome, it can't do its job

and VLCSFAs build up inside the cell. This build up is very harmful to nerve cells and causes the break down of the myelin sheath around these cells. Without the myelin, nerves are unable to transmit messages throughout the body resulting in symptoms of ALD.

Q6. How does ALD affect the function of the ER and the peroxisome within a cell?(A diagram might be more helpful than a written response.)

In the next step of this assignment you will simulate the role enzymes play in the metabolism of fatty acids in both a person with ALD and a healthy individual.

What's All The Fuss About Fats?

Part 3.

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Augusto's Paperclip model

Augusto Odone developed a model to understand the way our bodies make VLCSFAs. He used two kinds of paperclips to represent the two kinds bonds within fatty acids. Paperclips = carbon-carbon bond, chain of paperclips = fatty acid chain



Triangular paperclip = saturated bond

Rounded paperclip = unsaturated bond

Directions:

You will be working in groups of 2. Person 1 (taller) will act out the job of the enzyme in the ER and the Person 2 (shorter) will act out the job of the enzyme in the peroxisome. In this activity we will simulate fatty acid metabolism in a normal person, a person with ALD and a person with ALD receiving Lorenzo's Oil.

Scenario 1. Normal person

ER enzyme:

1. Take the diet cup and make long chain fatty acids by linking paperclips together in chains of 12.

Rules: Paperclip chains can only include one kind of paperclip (either all rounded or all triangular). You *must* alternate between making saturated and unsaturated fatty acid chains.

2. As chains are made, they are transported to the peroxisome to be broken down.

Peroxisome enzyme:

1. Take the chains given to you by the ER and break them down
2. Separate paperclips into piles

Record the total number of saturated and unsaturated fatty acids that the peroxisome enzyme **DID NOT** break down at the end of this scenario in the data table provided.

Q7. Describe your body's normal response to too many VLCsFA?

Scenario 2. ALD patient

Reset your diet cup with the paperclips separated by the peroxisome in the first scenario.

ER enzyme: Repeat the steps from scenario 1.

Peroxisome enzyme: A person with **ALD is unable to breakdown the long chain saturated fatty acids**; however, there are **other enzymes within the peroxisome that allow for the metabolism of unsaturated fats**.

1. Take the chains given to you by the ER. You are **NOT able** to break apart triangular paperclip chains (saturated fats). Set them aside.
2. You are able to break apart rounded paperclip chains (unsaturated fats). Break apart these chains and put paperclips in a pile.

Record the total number of saturated and unsaturated fatty acids that **DID NOT** break down at the end of this scenario in the data table provided.

Q8. What are the similarities and differences between a normal person and an ALD patient?

Q9. In your own words describe your role as the enzyme in either the ER or the peroxisome in the cell of a person with ALD.

Scenario 3. ALD patient treated with Lorenzo’s Oil

Get a new diet cup. This simulates the **diet with a reduced amount of saturated fats** as well as **addition of Lorenzo’s Oil**. **Lorenzo’s oil is a combination of two very long chain unsaturated fats.**

ER enzyme: Repeat the steps from scenario 1.

Peroxisome enzyme: Repeat steps from scenario 2.

Record the total number of saturated and unsaturated fatty acids that DID NOT break down at the end of this scenario in the data table provided.

Scenario	# non-metabolized Saturated fats	# non-metabolized Unsaturated fats
1. Normal person		
2. ALD patient		
3. ALD patient + Lorenzo’s Oil		

Q10. Use your data to compare and contrast the build-up of saturated fats in a person with ALD before and after treatment with Lorenzo's Oil and a modified diet. If you wish, use a Venn diagram for your response.

Q11. Now that you understand what Lorenzo's oil and the diet did for Lorenzo...

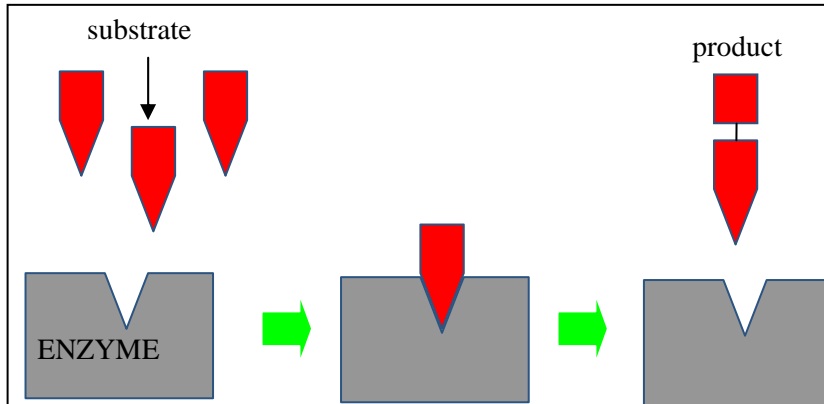
- *...would you make the same decision as Lorenzo's parents to give him "the oil" on your own without any medical approval? Explain.*
- *Do you think that what Lorenzo's parents did was "renegade" (outlaw) science? Explain.*

Part 4.

How Does Lorenzo's Oil Work?

Carefully study the diagrams below.

A



Q12. Which diagram shows Lorenzo's oil being used by an ALD patient's cells?

- ***Label the saturated and unsaturated fats on the diagram***

Q13. Explain how the diagram you chose in Q12 demonstrates why cells of ALD boys who are receiving Lorenzo's oil.

Q14. Is Lorenzo's oil a treatment or a cure. Explain and defend your answer with observations from the movie. (continue your response on the back if needed.)