Short Performance Assessment: **HS-LS1-1**

Grade Level: **High School**

Adapted from SNAP[1]

Title	**ALD Mutation**		
Designed by	**Steve Rhodes - American International School Dhaka**	Course(s)	**Biology**

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Performance Expectation	**HS-LS1-1**: Construct an explanation based on evidence for how the structure of DNA determines the structure of proteins which carry out the essential functions of life through systems of specialized cells. **Clarification Statement**: none **Assessment Boundary**: Assessment does not include identification of specific cell or tissue types, whole body systems, specific protein structures and functions, or the biochemistry of protein synthesis.

Science and Engineering Practice	**Construct Explanations** • Construct an explanation based on valid and reliable evidence obtained from a variety of sources (including students' own investigations, models, theories, simulations, peer review) and the assumption that theories and laws that describe the natural world operate today as they did in the past and will continue to do so in the future.
Disciplinary Core Ideas	**LS1.A: Structure and Function** • Systems of specialized cells within organisms help them perform the essential functions of life. • All cells contain genetic information in the form of DNA molecules. Genes are regions in the DNA that contain the instructions that code for the formation of proteins, which carry out most of the work of cells. (Note: This Disciplinary Core Idea is also addressed by HS-LS3-1.)
Crosscutting Concept	**Structure and Function** • Investigating or designing new systems or structures requires a detailed examination of the properties of different materials, the structures of different components, and connections of components to reveal its function and/or solve a problem

Student Performance	1. Articulating the explanation of phenomena 2. Evidence 3. Reasoning

[1] The Short Performance Assessment (SPA) and the Assessment Rubric adapted from the Stanford NGSS Assessment Project http://snapgse.stanford.edu/

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Name_____

Lorenzo's Oil
Genetics and Disease

Read the information below and answer the questions related to Lorenzo's oil.

Lorenzo suffered from a genetic disease which caused a mutation in his genes. Scientists have discovered that the *ABCD1* gene on the X chromosome contributes to people developing AlD. DNA is made out of the nucleotide sequences of four bases. Adenine, Cytosine, Thymine and Guanine. The *ABCD1* gene is comprised of over 300 bp of nucleotide. Here is a section of the *ABCD1* gene from a normal person and from a person who has AlD. Adrenoleukodystrophy. In people without the mutation for ALD, very long chained fatty acids are transported to peroxisomes inside their cells and broken down.

There are 65 different type of mutations that can cause ALD. Below is an example of a mutation which is occurring in protein transporter in a peroxisome.

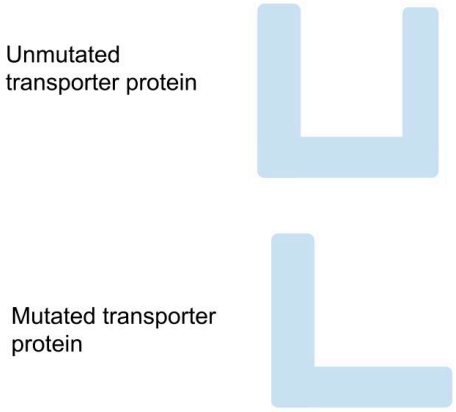
See the sequences below

Normal *ABCD1* Gene

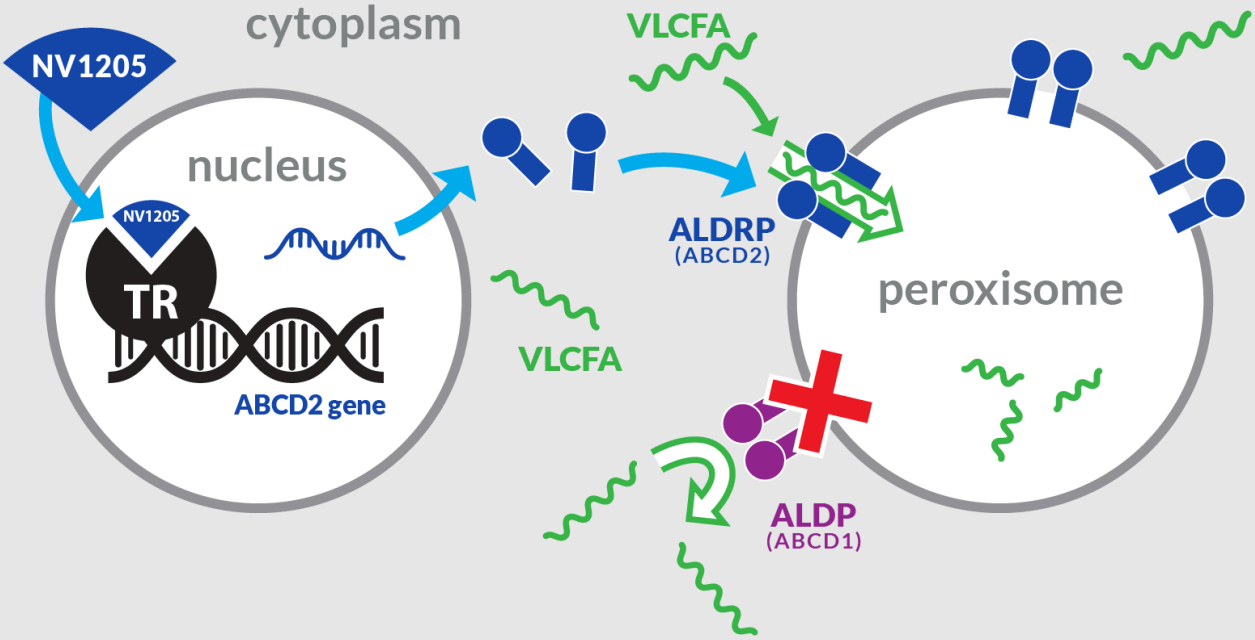
ATTGCCCGCCCCTTTAAATTTTGCCGCAAGCGGCTTA

Mutated *ABCD1* Gene

ATTGCCCGCCCCTTTAAATTATGCCGCAAGCGGCTTA



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Peroxisomes are small sacs within cells that process many types of molecules including VLCFA

ABCD1 gene: ATP-binding cascade subfamily D member 1 provides instructions for producing the adrenoleukodystrophy protein (ALDP).

ALDP normally facilitates degradation of VLCFA by the peroxisome; it is incapable of carrying out this function when mutated in X-ALD

VLCFA, or very long chain fatty acids, accumulate in certain cells of X-ALD patients and damage the cells.

ABCD2 gene: ATP Binding Cassette Subfamily D Member 2 provides instruction for producing the adrenoleukodystrophy-related protein (ALDRP).

ALDRP: ALD-related protein has similar function to ALDP and facilitates degradation of VLCFA by the peroxisome.

Degradation of VLCFA is restored, thereby reducing accumulation in cells

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Use all the information and data in the resource booklet to support your answer to the following question.

3. (a) How does the structure of DNA determine whether or not a person has ALD.

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Assessment Rubric* - Question 1				
	Emerging	Developing	Approaching Proficiency	Excelling
Description of performance				
Sample student responses				

Assessment Rubric* - Question 2				
	Emerging	Developing	Approaching Proficiency	Excelling
Description of performance				
Sample student responses				

Insert additional Assessment Rubrics (if needed) here.

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