**Woody Guthrie and Pedigree Analysis** 

1. Who was Woody Guthrie? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

2. What is his “claim to fame”? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

3. During the 1950’s he experienced times of very odd behavior during which he could not play the guitar and he often staggered and fell down. People assumed he was drunk. But he was not. What caused this odd behavior? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

4. On a separate piece of paper, create a pedigree of Woody Guthrie’s family based on the information from the timeline at your lab table. The following people in the pedigree are known to have had Huntington’s Disease – Nora(mom), Woody, Gwen, and Carolyn Sue. Note: Because the onset of the disease is later in life, others in the pedigree may have it, but have not yet shown symptoms or died before they showed symptoms.

* Shade in those known to have Huntington’s
* Draw a diagonal line through the symbols of those that are deceased
* Write the name of each individual and the date in which they were born/died

5. Is Huntington’s disease a dominant or recessive condition? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

6. Is the gene sex-linked or autosomal? How do you know from the pedigree? \_\_\_\_\_\_\_\_\_\_\_\_ \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

7. On which chromosome is the Huntingon’s disease (HD) gene found? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

8. Complete the punnett square that shows the probability of Woody’s parents having a child with Huntington’s. According to the results of the punnett square, what percentage of Nora and Charley’s children would have inherited the Huntington’s gene? \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_

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9. Huntington’s disease is caused by a repeat of the sequence CAG (3 of the 4 nucleotides found in DNA – **c**ytosine, **a**denine, **g**uanine, and **t**hymine) on the **coding strand** of DNA from chromosome #4. The number of repeated CAG sequences is directly related to the severity of the condition. A normal person has between 11 and 34 copies CAG in the Huntingtin gene. Examine the data tables below and answer questions 10 and 11.

**Table 1**

**Table 2**

table #1 from http://www.ninds.nih.gov/disorders/huntington/detail\_huntington.htm

|  |  |
| --- | --- |
| **No. of CAG repeats** | **Outcome** |
| < 28 | Normal range; individual will not develop HD |
| 29-34 | Individual will not develop HD but the next generation is at risk |
| 35-39 | Some, but not all, individuals in this range will develop HD; next generation is also at risk |
| > 40  | Individual will develop HD |

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| --- | --- |
| **CAG Repeat Size** | **Median Age at Onset \***  |
| **(years)****\*Age by which 50% of individuals will be affected.**Table #2 fromwww.compgene.com/hd.htm |
| 39 | 66 |
| 40 | 59 |
| 41 | 54 |
| 42 | 49 |
| 43 | 44 |
| 44 | 42 |
| 45 | 37 |
| 46 | 36 |
| 47 | 33 |
| 48 | 32 |
| 49 | 28 |

10. The Huntingtin protein is found in the cytoplasm of cells all over the body, is important for nerve and muscle function, and is known to be necessary for life. But the exact function is still unknown. The CAG repeat causes an abnormal form of the Huntingtin protein because the amino acid \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ is repeated, changing the structure and therefore, function of the protein. Research Huntington’s Disease to answer the question.

11. Complete a graph of the data in Table 2.



12. A number of people in the pedigree from #4 have an unknown genotype. What are the chances that Nora will get Huntington’s? \_\_\_\_\_\_\_\_\_\_\_\_\_\_

13. Nora decided not to be tested for the disease. Give reasons why she might have come to that decision. How might this decision affect her children?

14. Would you have done the same? Explain

15. Let’s say that Nora had a job as an air traffic controller at a major airport in the US. How might HD affect Nora’s ability to do her job?

16. Do you think the FAA or her employer should require that she be tested? Explain.

17. Nora’s sister Joady is 40 years old. She decided to be tested and was found to be positive for the defective gene. Joady is a pilot for an international airline, but plans to stay in her job and not inform her employer. The genetic counselor is torn between her legal responsibility of confidentiality and possibly being held legally responsible for failing to report Joady’s condition before a terrible plane accident occurs. What do you think the genetic counselor should do?