X-Linked Dominant Inheritance

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X-linked inheritance refers to the pattern of inheritance of a faulty (mutated) gene that is located on the X chromosome (X-linked genes). Males have one Y chromosome and one X chromosome and therefore only have one copy of the genes that make up the X-chromosome. Females have two copies of the X-chromosome genes (Female=XX, Male =XY). Shortly after conception, one of the X-chromosome copies in each female cell is randomly inactivated. This means that both males and females only have one active copy of the X-chromosome in each cell.

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If the body cannot work normally with less than the usual amount of correct gene product, a female will generally be affected by the X-linked mutated gene that she is "carrying". In these cases, the mutation making the gene copy faulty appears to override or "dominate" the unchanged information in the correct copy of the gene: it is described as a dominant mutation.

What is the risk of having a child affected by a X-Linked Dominant disorder?

Figure 1 illustrates the risk if a mother is affected by an X-linked dominant disorder. If a mother carries a Dominant mutation on one of her X chromosomes there is a 50% chance of passing the mutation on to each of her sons or daughters in each pregnancy. There is also a 50% chance that she will pass the normal X chromosome on and these boys and girls will be unaffected.

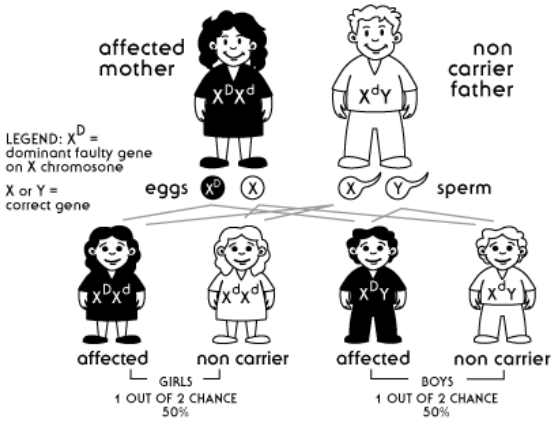
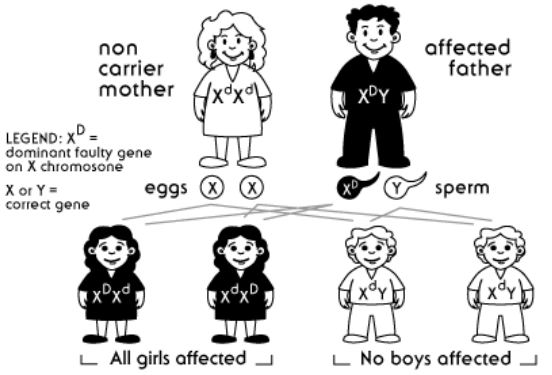


Figure 1: The faulty copy of the X-linked gene containing a dominant mutation is represented by "D". The correct X-linked gene copy is represented by "d". **(Picture from the Centre for Genetics Education website: http://genetics.com.au)**

Figure 2 illustrates the risk if the father is affected by a X-linked dominant disorder. When the father is affected by a condition due to an X-linked dominant mutation none of his sons can inherit the faulty gene since he only gives his Y chromosome to his sons. All of his daughters will be affected because they will receive his X-chromosome containing the faulty gene copy. Their unaffected mother will pass on only correct copies of the gene.



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