Your body is made up of trillions of tiny cells. Almost every cell in your body has a nucleus, a sort of headquarters that contains all our genetic information, or DNA.

DNA is packaged into structures called chromosomes. We have 23 chromosome pairs (for a total of 46 chromosomes) in each cell. Think of it like socks:

Mom contributes 1 chromosome for each pair and dad contributes 1 chromosome for each pair. Notice that the X and Y chromosomes which determine sex are the only pair of chromosomes which are not necessarily a matched pair. Women have two X chromosomes, but men have one X and one Y chromosome.

Each chromosome contains many genes. Genes are segments of DNA that determine specific human characteristics, such as height or hair color. Having two copies of each chromosome means we have two copies of every gene. Your mix of genes is unique to you. Even people in the same family have variations in their genes.

Think of the genome as a big instruction book telling your body how to grow and develop. Chromosomes are the chapters making up the book, and genes are sentences in each chapter.
**GENETICS CHEAT SHEET**

- There are 23 pairs of chromosomes in each cell of our bodies.
- Chromosomes contain sections of DNA called genes that tell our body how to grow and develop.
- We all have two copies of each chromosome and two copies of each gene.
- There is a 50% chance of inheriting an individual gene from a parent.
- Some genes are dominant. A single gene might be responsible for a certain function of the body.
- Some genes are recessive. Both gene copies work together to make a difference in the body.
- Changes in the "spelling" of a gene (mutations) are mostly harmless but sometimes cause problems.

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**What is von Hippel-Lindau**

Von Hippel-Lindau (VHL) is a condition associated with the VHL gene which is located on chromosome 3. Everyone has this VHL gene, but people who have a mutation or variation in this gene have an increased risk of developing clear cell renal cell carcinoma, a type of kidney cancer. People with VHL also have an increased risk of developing a pheochromocytoma, a tumor of the adrenal gland, as well as blood vessel tumors of the brain, spinal cord, inner ear, and eye. VHL can also affect the lungs, liver, pancreas, and reproductive organs.

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**How is VHL Inherited?**

Normally, every cell has two working copies of each gene: one inherited from the mother and one inherited from the father. Some genetic conditions have **recessive** inheritance and are caused by two improperly working copies of a gene.
VHL follows a dominant inheritance pattern, meaning that VHL is caused by having one improperly working copy of the VHL gene in each cell. Someone who has VHL will have one normal copy of the gene and one copy with a mutation. This means that an affected parent has a 50% chance to pass on the normal copy of the gene to a child and 50% chance to pass on the mutated copy, in which case the child would also be affected with VHL. There is nothing anyone can do or not do to control which gene is passed on; it is completely up to chance. Therefore, a child who has a parent with a mutation has a 50% chance of inheriting that mutation.

More family planning information is available in the VHL Handbook (Adult Issues and Family Planning).
How Common is VHL?

It is estimated that about 1 in 36,000 people has VHL. Approximately 80% of patients with VHL inherit the VHL mutation from a parent who is also affected with VHL. The remaining 20% of people affected with VHL do not have any family history of the condition. This is because instead of inheriting the VHL gene mutation from a parent, the individual has a ‘new’ mutation that occurred randomly in the VHL gene during embryo development. These individuals are the only person in their family to have VHL but they now have a 50% chance of passing the VHL mutation on to each of their children.

Where Can I Learn More?

Here is a good starting list of websites to learn more about VHL:

- VHL Alliance (vhl.org)
- Genetics Home Reference
- The Children’s Hospital of Philadelphia (CHOP)
- Cancer.Net
- UptoDate*

There are a lot of other people and places you can go to for further information. Talking to family members, doctors, peers, and teachers can be helpful in understanding and dealing with VHL. Some resources might be more helpful than others depending on whether you are trying to learn facts about your health, talk about your feelings, or learn about what support is available. There are pros and cons for any educational resource; here are a few to keep in mind for some of the sources of information.

<table>
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<tr>
<th></th>
<th>Pros</th>
<th>Cons</th>
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<tbody>
<tr>
<td>The Internet</td>
<td>There is a lot of information out there and you can look it up when it is convenient for you.</td>
<td>The amount of information available can be overwhelming. And it can be hard to know what is accurate.</td>
</tr>
<tr>
<td>Family &amp; Friends</td>
<td>Your family and friends may love you a lot and know you very well. You may also have family members that know what it is like to have VHL.</td>
<td>Family and friends might not have the most accurate information. You also may not feel comfortable talking with them about “adult” topics.</td>
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<tr>
<td>Doctors</td>
<td>Doctors have accurate information about your health. VHL specialists will also be knowledgeable about VHL.</td>
<td>Non-specialists may not know very much about VHL. Doctors often have limited time, and they may not understand your life outside the doctor's office.</td>
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