Effectiveness of Hydroxyurea and Blood Transfusions to Treat Sickle Cell Disease

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Abstract

Sickle cell disease is a hereditary disease that has a series of problems, the most common one being pain episodes. Currently the two most common treatments for sickle cell disease is hydroxyurea and blood transfusions. Both treatments have negative effects but none of those effects are important enough to stop the use of the treatments. There has also not been an actual statistical comparison between the two treatments, so it is unknown which one is more effective than the other. This paper analyses the two different treatments and focuses on three different factors of the disease, the amount of acute chest syndrome evens, painful crisis events, and infections. All three symptoms lower the quality of life of the SCD carrier by interfering with their everyday tasks and forces them to live in a specific way in order to reduce the amounts of negative events.

Introduction

Sickle cell disease (SCD) occurs in approximately 80,000 people in the United States each year (Centers for Disease Control and Prevention, 2017). SCD is a group of hereditary red blood cell disorders caused by the sickle shaped cells (Figure 1), which form due to problems with hemoglobin, a protein that helps carry oxygen in red blood cells through the bloodstream. When the hemoglobin in a red blood cell arrange into long and parallel fibres, stretching out the cell, it is known as polymerization, which is the cause of SCD. The symptoms of SCD include anemia, pain due to blocked blood vessels, acute chest syndrome, vaso occlusive crisis, an increase in infections, swelling of hands and feet, and in some cases, vision problems. Currently, hydroxyurea, an oral medicine taken daily to prevent the adverse effects of SCD, is the main drug used as a treatment for SCD. Hydroxyurea increases the amount of fetal hemoglobin (HbF) in the blood, providing protection against HbS, since it inhibits the HbS polymerization. Another method used to treat SCD is blood transfusions which increases the amount of normal red blood cells, because they can live longer in the circulatory system and have a larger surface area than red blood cells with sickle hemoglobin. Blood transfusions also decrease the risk of clogged blood vessels thus improving oxygen delivery throughout the body.

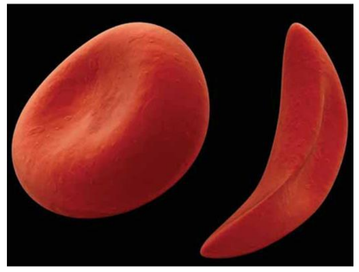

Figure 1. Normal RBC vs. Sickle cell

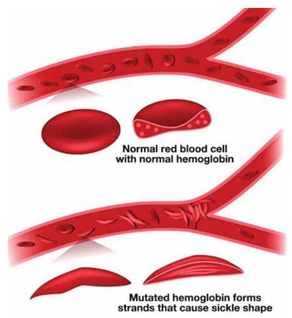

Figure 2. Flow of RBC in blood vessel vs. flow of sickle cell

Methods and Materials

The design of this paper is that of a systematic literature review. Research databases including Google Scholar, PUBMED-NCBI, ResearchGate, ScienceDirect, Blood Journal, PLOS, BioMed Central, etc. were used to gather studies analyzing the effectiveness of hydroxyurea and blood transfusions as treatments for sickle cell disease. Keywords, such as “sickle cell disease,” “hydroxyurea treatment,” “blood transfusions,” etc. were used to collect informative articles. Additional information was gathered from the references of used articles. Using a systematic literature review was the most effective method for this specific study because conducting experiments would not be needed to gather data as both treatments already have a significant amount of literature to support their respective costs and benefits. It also would not be possible to conduct such experiments due to the constraints of a high school setting. Other methods such as using secondary data or surveys are not appropriate either. Secondary data would not provide enough information to complete the data collection and surveys do not provide the same type of data that is needed.

Discussion

When comparing the results of the three different events of SCD, both infections and painful crisis saw greater differences in the rate of occurrences than acute chest syndrome when compared between hydroxyurea and blood transfusion. All three of the average percentages of the events were greater for hydroxyurea, meaning that there was a higher rate of symptoms among the patients who used hydroxyurea than those who had blood transfusions. Patients that received hydroxyurea as their treatment saw a higher chance of having an infection. The p-value for acute chest syndrome comparing hydroxyurea with blood transfusions was 0.24 and the p-value for painful crisis was 0.39. The p-value for infections was 0.16 which is lower than both of the other two, however, all three values are greater than .05 meaning that the alternative hypothesis is rejected and the null hypothesis is accepted. Through analysis of the three factors, acute chest syndrome, pain crisis, and infections, the p-values reflect that there is no significant difference in effectiveness between blood transfusions and hydroxyurea when treating SCD.

Conclusions

The systematic literature review conducted on evaluating the effectiveness of hydroxyurea and blood transfusions as treatments for SCD shows that the alternative hypothesis should be rejected and that the null hypothesis should be accepted. Both treatments are currently being used widely for SCD patients and although their effects on acute chest syndrome and vaso occlusive crisis are similar, hydroxyurea has shown to have higher rates of infections among the patients. There are precautions before receiving a blood transfusions which can help lower the chance of an infection, however, it is much harder to lower the risk of infections from hydroxyurea since the amount of white blood cells and platelets are being lowered. A temporary decrease in platelets can also lead to anemia and an increase in bleeding, both of which are not seen in blood transfusions. Although quantitatively, the data suggests that there is not a significant difference between the treatments, qualitatively, there is a difference.

Results

	Number of Patients	Acute chest syndrome	Painful Crisis	Infection
Alvarez, O. (2013)	66	4	62	3
DeBaun, M. R. (2014)	99	5	96	1
Miller, S. T. (2001)	59	2	9	-
Miller, S. T. (2001)	63	4	17	-
Vichinsky, E. P. (2000)	264	16	47	3
Wang, W. C. (STOP 1998) (2013)	63	4	11	0
Ware, R. E. (2016)	61	3	23	4
Average	96	5.4	37.9	2.2

	Number of Patients	Acute Chest Syndrome	Painful Crisis	Infections
Alvarez, O. (2013)	67	9	38	9
Gulbis, B. (2005)	32	3	22	0
Gulbis, B. (2005)	92	3	31	0
Jain, D. L. (2012)	30	0	18	10
Kinney T. R. (1999)	84	10	76	23
Koren, A. (1999)	60	7	39	-
Wang, W. C. (2013)	96	8	63	0
Ware, R. E. (2016)	60	5	11	4

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Acknowledgements

I would like to thank Dr. Nikki Malhotra for all her time spent helping me with my research during every part of my research process. Additionally, I would like to thank my expert advisors Dr. Gregory Kato for helping me most during the beginning of my project, narrowing down my research project, and also Dr. Martin Steinberg for his support throughout my research process and help with any questions about my topic. Lastly, I would like to thank Kea Turqueza for helping me with my research and giving me advice based off of her experience with AP research.