Chapter 7
Developing a National Registry for Hemochromatosis

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ABSTRACT

Hereditary Hemochromatosis (HH) is a disorder where iron and ferritin concentrations in a patient's blood are much higher than normal healthy levels. The main therapeutic intervention for individuals with HH is removing 300-500 mL of blood every few months to maintain ferritin concentration within acceptable ranges. The blood collected during these venesections is usually discarded as there is a belief that blood with high levels of ferritin are not suitable for blood transfusion purposes. Australian Red Cross Blood Services voluntarily collects blood from donors for subsequent use in blood transfusion. Annually more than 700 thousand units are transfused within Australia and there is a constant need for new donors given the significant imbalance between supply and demand of blood products. Besides red cell transfusions, the Red Cross also issues donor blood for development of many other blood products essential for patient health care. The HH blood can currently be used for other blood products if not for red cell transfusion. However, there is evidence to suggest that there is no significant difference between the red cells of the normal healthy population compared to those from HH patients. Australian Red Cross has developed a mobile computer application (High Ferritin "app") as they have started collecting blood from HH patients. Though there is little or no awareness about the existence and use of this High Ferritin app in general HH population, their doctors and nurses collecting their blood for therapeutic purposes. This chapter describes possibility of saving and utilizing the blood collected from hemochromatosis patients for therapeutic purposes. A national hemochromatosis patients registry, in collaboration with High Ferritin app (HFa) developed by Australian Red Cross Blood Services, accessible to the patients, their doctors and Red Cross Blood Collection Sservices 24 hours a day anywhere in the country can allow the patients to donate the blood collected for therapeutic purposes at any affiliated blood collection center in the country after they automatically get a message either by email or text message after their blood results have been reviewed by their doctor and they are required to go for venesection.

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INTRODUCTION

Hereditary Hemochromatosis (HH) is an inherited disorder of iron metabolism resulting in gradual accumulation of iron (Franchini, 2006; Janssen & Swinkels, 2009; Limdi & Crampton, 2004). As there is no physiological method to remove iron from the body, save for its loss in faeces and through the sloughing of epithelial cells, abnormal iron uptake results in eventual iron overload. This excess iron is deposited in essential organs such as liver and kidney, leading to secondary cirrhosis and diabetes (Andrews, 1999; Franchini, 2006; Janssen & Swinkels, 2009). One of the most common inherited disorders in the Caucasian population, HH is autosomal recessive, the prevalence of which is roughly 1 in 500 (Limdi & Crampton, 2004). Its effects are gradual, and affected individuals only begin to show symptoms in the third to fourth decades of life including hepatomegaly, fatigue, and skin pigmentation (Andrews, 1999; Limdi & Crampton, 2004).

The most economic form of treatment for individuals with HH is therapeutic phlebotomy or venesection (Janssen & Swinkels, 2009). This frequent removal of 450-500 mL (1 unit) of blood is implemented with the primary goal of decreasing/maintaining iron concentrations within normal ranges (Janssen & Swinkels, 2009; Limdi & Crampton, 2004). The blood collected during venesection is typically discarded at collection clinics as there are concerns about its utilization for transfusion purposes (De Buck, Pauwels, Dieltjens, Compernolle, & Vandekerckhove, 2012; Janssen & Swinkels, 2009). The two most common arguments for discarding blood collected from HH donors are focused on its safety and the voluntary status of HH donors, as they too benefit from the donation (De Buck et al., 2012). Given the value of blood and the increasing need of it in today's society, however, it is plausible that these arguments require further investigation.

This chapter discusses the possibility of utilizing the blood collected during routine venesection of HH patients for various therapeutic purposes. A national hemochromatosis patients registry, in tandem with the High Ferritin app (HFa) developed by Australian Red Cross Blood Services, would facilitate matching HH patient venesections with transfusion requirements and therefore may address the current imbalance between blood supply and demand.

BACKGROUND

The Potential of Cell Salvage Programs

Transfusion medicine is a clinically oriented practice with an emphasis on patient care (). Clinicians now have a variety of blood products that may be administered to patients as required, including whole blood, packed red cells, fresh frozen plasma and platelets, among others (). Packed red blood cells is the most commonly transfused blood component, where approximately 75 million units are collected and administered annually worldwide (). In 2013 703,358 red cells UNITS? were issued for transfusion within Australia (Australian Red Cross Annual Report 2013-14). However issues arise in terms of safety and availability of allogenic blood stocks (). The former is beyond the scope of this review; however the latter can be addressed by the use of HH blood.

Leitman et al conducted an analysis over a 27 month period where donations from HH patients resulted in a significant increase in their allogenic inventory. These authors demonstrated WHAT TYPE? benefits to HH donors who expressed personal satisfaction in knowing their blood would be of use rather