PP 70. Sickle Cell Disease (SCD): A Case Study

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Background
In November 2013, a sample from patient X, diagnosed with Sickle Cell Disease (SCD) prior to her pregnancy, was referred to Reference Laboratory for investigation. The patient had been previously transfused with no complications; however, problems were encountered upon subsequent requests. SCD, an inherited haemoglobinopathy, raises much concern as patients often present with serological incompatibilities due to repeated blood transfusion serving as the cornerstone of treatment. Depending on the serological picture, the appropriate interventions and handling of these requests in terms of transfusion therapy needs to be exercised.

Methods
Initial antibody screens and compatibility testing by the Indirect Antiglobulin Technique (IAT) were conducted in the bloodbank as per the standard protocol. Reference Laboratory performed antibody identification by the Saline Immediate Spin, IAT and gel column agglutination methods against the standard panel along with a range of rare cells. Current protocols regarding selection of blood for SCD patients suggest leuco-depleted red blood cells phenotypically matched for Rh and Kell specificity which was performed by tube and gel methods.

Results
Compatible units were not found by the bloodbank. Testing by the Reference Laboratory identified anti-U by saline IAT as all standard panels and rare cells were found positive in the antibody investigation, with the exception of U negative cells. Mixed field results were obtained during typing of the patient's cells and the red cell phenotyping was therefore inconclusive. The patient's red cells were presumed U negative due to the presence of the anti-U antibody. In addition, the patient's cells were found to be sensitized with IgG antibodies.

Conclusion
This case demonstrates the need to manage the ongoing requests for blood for cases of Sickle Cell Anaemia correctly. Complications that arise are not only due to alloantibody formation but also due to the limitations caused by sensitization and post transfusion specimens. Molecular genotyping serves to eliminate the restrictions caused by routine phenotyping (tube/gel) and renders more conclusive results. Treating individuals that present with rare blood types is challenging. It is therefore necessary to inform the attending clinicians to notify the laboratory timeously of requests for blood in order to avoid delays. Where fresh units are unavailable or cannot be procured, rare units in frozen storage will have to be thawed. In cases where the request cannot be met due to limited stocks, the Monocyte Mononuclear Assay, currently under validation in our laboratory, may be beneficial. This enables the analysis of the in vivo red cell survival of incompatible units through in vitro testing, thus assessing the potential risk of a haemolytic transfusion reaction. However, communication is vital and efforts must be made to enhance rare blood stock levels.