

# Your hereditary RBC defect workflow streamlined – based on the latest research

Abnormal RBC parameters are challenges in the daily lab routine. Moreover, the need and high interest for reliable diagnosis of haemoglobinopathies and red cell membrane disorders from other causes of abnormalities in RBC indices have been expressed frequently. Recent research addressed these challenges and offers an opportunity to streamline the laboratory workflow and choose the appropriate confirmatory tests.

## Your benefits in daily routine

- Free your hands with less manual work
- Save precious time and optimise TAT
- Streamline your laboratory workflow for abnormal RBC samples

#### A valuable tool you can rely on

- Confident handling of suspicious RBC disease samples
- Support the identification of hereditary RBC diseases, even when anaemia is absent
- Choose the appropriate confirmatory test to confirm the clinical root cause straight away



# **RBC Defect Workflow Optimisation in Extended IPU**

Based on the research of Nivaggioni *et al.* and Berda-Haddad *et al.*, Sysmex introduces the RBC Defect Workflow Optimisation (RWO), a new rule set implemented in the *Extended* Information Processing Unit (IPU). This rule set combines the CBC-O application, the RBC score and advanced RBC and RET parameters. The intelligent two-step algorithm can be used to support the identification of patients with hereditary RBC diseases and iron deficiency anaemia (IDA). Improved laboratory workflow is achieved by forwarding suspected samples for hereditary RBC diseases to the respective tests that can confirm the diagnosis and at the same time correctly exclude IDA samples that can generate spurious results.

Abbreviations: RBC – Red blood cells, HGB – Haemoglobin, HCT – Haematocrit, HYPO-He – Percentage of hyperchromic red blood cells, IRF – Immature reticulocyte fraction, MCHC – Mean corpuscular haemoglobin content, MCH – Mean corpuscular haemoglobin, MCV – Mean corpuscular volume, MicroR – Percentage of microcytic red blood cells, NRBC – Nucleated red blood cells, RDW-SD – Red blood cell distribution width-standard deviation. RBC score: calculated value based on FRC% (percentage of fragmented red blood cells) and RET# (reticulocyte count).

# RBC DEFECT WORKFLOW OPTIMISATION

Know more. Decide with confidence. Act faster.



#### Requirements

The implementation of RWO requires a Sysmex automated haematology analyser equipped with a RET channel and connected to the work area manager *Extended* IPU.

#### **Overview of essential parameters and applications**

Diagnostic CBC RBC, HGB, HCT, MCHC, MCH, MCV, RDW-SD, MicroR, NRBC Diagnostic RET RET#, IRF, HYPO-He Research RET FRC%, RBC-O, HGB-O



**Extended IPU** CBC-O application Rule set for RWO

### **Results from the publications**

Nivaggioni *et al.*, in a series of two studies [1, 2], developed and evaluated a reliable classification workflow for RBC diseases, both hereditary (including heterozygous haemoglobinopathies, sickle cell disease, hereditary spherocytosis and Southeast Asian ovalocytosis) and acquired (iron deficiency anaemia). The workflow includes the CBC-O application and RBC score described in the Berda-Haddad *et al.* study [3] for cases with truly elevated MCHC (> 36.5 g/dL, 22.7 mmol/L), along with several decision steps for samples with both normal and elevated MCHC. According to the publication, the decision tree reached a classification rate of 99.4 %, and had a sensitivity of 95.2 % and a specificity of 99.9 % for identifying RBC diseases in the studied cohort.

## Confirmatory workflow in the laboratory

After the identification of the respective sample with RBC Defect Workflow Optimisation rules in *Extended* IPU, the confirmatory test of the likely disease can be directly performed. This saves precious time and resources in the lab.



#### RBC diseases part of the RBC Defect Workflow Optimisation

#### Hereditary spherocytosis (HS)

Inherited red cell membrane disorder characterised by the presence of spherical red blood cells that have lost their central concavity. Defects in membrane proteins alter the physiology of the cells, with spherocytes losing their deformability and becoming trapped as the blood is being filtered in the spleen. Though some spherocytes escape and re-enter the circulation, they can still be identified with laboratory tests.

#### Southeast Asian ovalocytosis (SAO)

Inherited red cell membrane disorder arising from a mutation in the band 3 transmembrane protein that causes a dysregulation of the ionic exchange and structural abnormalities. The red blood cells acquire a distinctive oval shape which affects the deformability, but individuals with this disease do not present with severe clinical symptoms, apart from signs of mild haemolysis and jaundice.

Heterozygous haemoglobinopathies

Large group of genetic disorders describing

quantitative or qualitative disturbance in

the production of the haemoglobin protein

chains. The main adult haemoglobin molecule

comprises of two  $\alpha$ -globin and two  $\beta$ -globin

chains  $(\alpha_2\beta_2)$ . Thalassaemias, a large group

mutations in the alpha or beta chain of hae-

moglobin that lead to reduced production or

of haemoglobinopathies, are caused by

absence of the respective chain.

(HGB HTZ)

#### Sickle cell disease (SCD)

Inherited disease caused by mutation in the  $\beta$ -globin gene of haemoglobin A which results in the generation of haemoglobin S or HbS. Under low oxygen level conditions, the red blood cells acquire a sickle shape, causing ischaemic episodes as they block the blood flow in capillaries. Sickle cells have a very short lifespan, up to 20 days, that contributes in a rate of haemolysis that the bone marrow cannot compensate.

#### Benefit from more background information in our freely accessible white papers: www.sysmex-europe.com/whitepapers

# References [1] Nivaggioni V et al. (2020): Int J Lab Hematol. 42(6): 697–704. [2] Nivaggioni V et al. (2022): Int J Lab Hematol. 44(2) 84–86. [3] Berda-Haddad Y et al.(2017): Int J Lab Hematol. 39(1): 32–41.

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The rule set of the RBC Defect Workflow Optimisation in Extended IPU was developed by a key opinion leader as a result of the mentioned research studies and is not CE marked.

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