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Rare anaemias, sickle-cell disease and COVID-19

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Summary. For rare haematological diseases (RHD), the first question to be answered is if patients with benign red blood cell (RBC) defects like haemoglobinopathies, membranopathies and enzymopathies are more vulnerable to COVID-19 infection. Up to now, there is no yet literature on the subject, but, like in general population, the presence of comorbidities such as diabetes, heart disease, pulmonary hypertension, reduced kidney and/or liver function, worsen the effects of the infection. Splenectomy may be an additional risk factor. (www.actabiomedica.it)

Key words: Rare anemias, sickle cell disease, coronavirus infection

Coronavirus disease (COVID -19) is due to a virus of unknown origin, that has spread from the Wuhan province in China, and that has become a global pandemic affecting about 170 countries. Currently, the epicenter for Europe is Italy, for America is USA, and for South West Asia it is Iran, The most recent WHO statistics for the world indicate a total of 2..074.529 confirmed cases and 139.378 confirmed deaths, with an estimated mortality of about 5%.

COVID-19, like SARS of 2002-3, is transmitted from the human carriers through respiratory droplets and affects primarily the pulmonary tract leading to severe respiratory insufficiency and dead, especially in aged men. At worldwide level, in order to keep people safe from coronavirus, the WHO has launched a dedicated message in Arabic, French and Spanish with partners WhatsApp and Facebook that with a capacity to reach 2 billion people enables WHO to get information directly into the hands of the people that need it (1).

In Europe, with the aim to support clinicians in hospitals that are currently facing the coronavirus emergency, on 24 March, the European Commission (EC), based on the experience with the European

Reference Networks (ERNs), launched the "COVID-19 Clinical Management Support System" (2). Ultimately, it is in the vital interest of the patients infected with COVID-19 that their doctors can discuss their cases, and get the best advice possible**.** This is so because many patients are in need of highly specialised care and, unfortunately, the practical experience on how to manage patients, in particular severe cases, is scarce and scattered in Europe (3).

For rare hematological diseases (RHD), the first question to be answered is if patients with hemoglobinopathies, like thalassemia and **sickle-cell disease (SCD),** are more vulnerable to COVID-19 infection. Hemoglobin disorders are generally not associated with respiratory conditions; however, com- plications involving the heart, lungs and the immune system, can be present in these patients, and, as in a SARS-CoV-2 positive patient, may trigger very serious complications. According to the most recent **EuroBloodNet report**, there is no yet literature on the subject, but some indications can be given from the experience with previous coronaviruses infections leading to respiratory infection (4). Like in general population, the presence of comorbidities such as diabetes, heart disease, pulmonary hypertension, reduced kidney and / or liver function, worsen the effects of the infection.

As mentioned before, among the patients with RHD, one group particularly at risk is SCD. As well

known, one serious complications and major cause of death in SCD is the acute chest syndrome (ACS), which is most often triggered by a respiratory infection. As demonstrated in USA, during the two influenza seasons (2003-2005), children with SCD were hospitalized 56 times more frequently than children without SCD, and rates were twice as high compared to children with cystic fibrosis. Accordingly, it seems that the SARS-CoV-2 infection may trigger such a serious complication and require special alertness on behalf of physicians treating infected patients. In addition, hypoxia, dehydration or acidosis due to respiratory infection may trigger a vaso-occlusive crisis and the ACS, the main cause of morbidity in SCD, frequently triggered by infectious events. Measures to prevent and treat ACS, early in the event of viral infection, must, therefore, be seriously considered. Moreover, since many SCD patients are treated with hydroxycar bamide (hydroxyurea), a cytotoxic agent, with possible immune-compromising effects, an increase in the severity of known viral infections may also contribute to an adverse outcome of these patients. A very complete haemoglobinopathy Patient Care Pathway suggesting changes to be made during the outbreak of COVID-19 in the admittance of patients to haemoglobinopathy day care centres /units /clinics has been published by **Thalassemia International Federation** (5).

Concerning **splenectomy** or functional hyposplenism, it do not represent a risk for a viral infection itself, but since SCD patients are more vulnerable to bacterial infections, it is important to prevent or to treat any bacterial superinfection. Concerning patients with **other rare anaemias**, they are at higher risk of COVID-19 and should be socially isolated. Specific guidelines on the management of these patients have been published in the UK Haemoglobinopathy Coordinating Centres (6).

Finally, one important aspect of patients with RHD is the **effect of blood transfusion** and the possibility of COVID-19 transmission through donated blood. Up to now there is no evidence of those conditions, such as pyruvate kinase deficiency, will be exacerbated by COVID-19, however patients should follow national guidelines for blood donation, and avoid reluctance due to infection.

Conflict of interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

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