Since January 2020 Elsevier has created a COVID-19 resource centre with free information in English and Mandarin on the novel coronavirus COVID-19. The COVID-19 resource centre is hosted on Elsevier Connect, the company's public news and information website.

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Letters to the Editor

Why continued lipoprotein apheresis is vital for homozygous familial hypercholesterolemia patients with COVID-19

Current expert opinion emphasizes that homozygous familial hypercholesterolemia (HoFH) patients with COVID-19 need to continue regular lipoprotein apheresis (LA).[1] LA is vital to prevent the progression of atherosclerotic cardiovascular disease (ASCVD) starting at very young ages in such patients.[2] A recent study from Turkey showed that access to LA deteriorated during the current pandemic thus exerting a strong negative impact on the chronic treatment of a highly vulnerable patient population being at increased risk of COVID-19 mortality.[3] Thus, the COVID-19 pandemic resulted in the cessation of a vital treatment even among patients who are aware of the severity of their genetic condition if left untreated.

Serum low-density lipoprotein cholesterol level (LDL-C) levels are four-to five-fold elevated in HoFH resulting in the very early onset of ASCVD and even fatal myocardial infarction in early childhood.[4] In addition to the ever-increasing cholesterol burden, HoFH patients often have an elevated serum lipoprotein(a) [Lp(a)] level, presenting another genetic causal ASCVD risk factor.[5] Jointly, elevated LDL-C and Lp(a) levels cause endothelial dysfunction.[6,7] Furthermore, the proinflammatory and antifibrinolytic features of serum Lp(a) contribute to the thrombogenic mechanisms of ASCVD.[8]

COVID-19 can be considered an endothelial disease that is particularly harmful in HoFH patients with preexisting dysfunctional endothelium.[9-11] In HoFH patients with COVID-19, the burden of endothelial injury is increased further by the cytokine storm and viral infection of the endothelial cells, so leading to an increased tendency to thrombosis both in the micro- and macrovasculature. In hospitalized COVID-19 adult patients in general, acute myocardial infarction occurred in 3.3% (95% CI 0.3–8.5) of cases.[12] It can be assumed that the event rate will be even higher among hospitalized HoFH patients.

LA acutely reduces serum LDL-C and Lp(a) levels by up to 70%, but the levels rapidly increase so requiring continuously repeated LA sessions.[13,14] Importantly, in HoFH patients, LA improves endothelial function.[15,16] However, because HoFH patients with SARS-CoV-2 infection appear to be especially vulnerable to thrombotic events, it is important to consider intensification of the LDL-cholesterol-lowering pharmacotherapy by adding a PCSK9 inhibitor to the normally required high-intensity statin treatment.[17] In addition to the effective decrease in LDL-C level, PCSK9 inhibition may enhance the antiviral action of interferon.[18] Overall, the clinicians treating HoFH children with COVID-19 need to pay very special attention to the continuation of LA and efficient lipid-lowering pharmacotherapy.

Conflict of interest

FJR has received research grants, honoraria, or consulting fees for professional input and/or delivered lectures from Sanofi, Regeneron, Amgen, and Novartis. RK as head of Apheresis Research Institute received research grants from Diamed, Cologne, Germany, and Asahi Kasei Medical, Tokyo Japan. PTK has received consultancy fees, lecture honoraria, and/or travel fees from Amgen, Novartis, Raisio Group, and Sanofi.

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Collateral damage of the COVID-19 pandemic on the management of homozygous familial hypercholesterolemia

With our 20-year of experience on lipoprotein apheresis (LA),[1,2] we fully agree with Vuorio et al's comment[3] regarding the vital role of continued LA in homozygous familial hypercholesterolemia (HoFH) patients during the COVID-19 pandemic. Our recent survey[4] was conducted to determine the collateral damage that the pandemic caused to provide insight to the impact of COVID-19 outbreak on lifestyle, anxiety levels, and management of HoFH in terms of LA and lipid-lowering therapy (LLT). The results of our descriptive survey depicted that management of patients with HoFH with regard to heart-healthy lifestyle and access to LA has significantly deteriorated during the pandemic even in a country where LA is full reimbursed and widely available.[4,5] Our study revealed that although LA was accessible in most of the centers throughout the country during the pandemic, most of the patients (75%) were not attending their LA sessions due to fear of contracting coronavirus.[4]

LA is not only the most effective means of low-density lipoprotein cholesterol (LDL-C) lowering, but also effectively decreases the number of inflammatory proteins including cytokines and thrombogenic factors and improves endothelial function.[2,6,7] LA also has impact on gene translation and transcription by affecting cytokine receptor functions.[8] Therefore, apheresis is also used as a therapeutic modality in patients with a cytokine release storm during the course of COVID-19.[9] However, LA is a hospital or healthcare center dependent treatment that can be easily disrupted by a disaster as we experienced during the COVID-19 pandemic either due to the overload of the medical system by COVID-19 patients or the reluctance of the patients to visit the hospital in fear of getting infected during the procedures.[1,4] Even patients with acute coronary syndromes have avoided admitting to hospital during the pandemic as several reports have shown all over the world.[10,11] We demonstrated that many HoFH patients could not get access to LA therapy during the pandemic exposing them to increased risk of cardiovascular events.[4] It is well established that the on-treatment LDL-C level is the major determinant of survival in HoFH.[12] Therefore, we emphasize in our paper the importance of continued regular effective therapy. Moreover, as we mentioned in our manuscript the role of National Scientific Societies and, more importantly, patient organizations is vital for increasing the awareness of