PRESS RELEASE

Case reports on lipoprotein-X hyperlipidaemia in Chinese paediatric patients with liver graft-versus-host disease post-hematopoietic stem cell transplantation

(Hong Kong, 20 February 2023) – **Graft-versus-host disease (GVHD) of the liver is one of the known complications of allogeneic hematopoietic stem cell transplantation (HSCT). Severe hypercholesterolaemia (a type of hyperlipidaemia) mediated by lipoprotein-X (Lp-X) in patients with liver GVHD who have underwent allogeneic HSCT is a recognised yet overlooked phenomenon. Reports in literature is limited in both adult and paediatric population. A group of doctors from Hong Kong Children's Hospital and Queen Mary Hospital reported two cases to look deeper into the issue. To the best knowledge of the authors, this is the first report in Chinese children.**

Lipoprotein-X is a major cause of hyperlipidaemia in cholestasis when bile constituents reflux from the bile ducts or hepatocytes to the blood stream. Unlike low-density lipoprotein (LDL-C; often called the 'bad' cholesterol), as Lp-X does not contain apolipoprotein B, the major component of LDL and one of the most important factors in the pathogenesis of atherosclerotic plaques, it is not atherogenic. Besides, owing to the same reason, Lp-X cannot be internalised into hepatocytes as the apolipoprotein is the most important ligand to the hepatic LDL-C receptor. Since Lp-X hypercholesterolemia is not due to overproduction by hepatocytes, use of medications such as statins to downregulate cholesterol synthesis is ineffective and unnecessary.

The medical team discussed two local cases, who were Chinese children underwent HSCT. The team hopes that transplant physicians and endocrinologists can be more aware of the association of severe hypercholesterolaemia mediated by Lp-X in post-haematopoietic stem cell transplantation patients with liver GVHD and avoid unnecessary and ineffective usage of statins.

The case report "Lipoprotein-X hyperlipidaemia in Chinese paediatric patients with liver graft-versus-host disease post-hematopoietic stem cell transplantation: two case reports" was published in the *Hong Kong Medical Journal*. https://doi.org/10.12809/hkmj219765

新聞稿

造血幹細胞移植後肝移植物抗宿主病中國籍兒童患者的脂蛋白-X 高血脂症病例報告

（香港，2023 年 2 月 20 日）— 肝臟的移植物抗宿主病是異體造血幹細胞移植的已知併發症之一。在接受異體造血幹細胞移植的患者中，肝臟移植物抗宿主病患者患上由脂蛋白-X 導致的嚴重高膽固醇血症（其中一種高血脂症）是一種公認但被忽視的現象。不論成人或兒童，報告這種病例的文獻有限。來自香港兒童醫院及瑪麗醫院的醫生發表了相信是首兩宗中國籍兒童病例報告，以更深入了解有關情況。

當膽汁成分從膽管或肝細胞回流到血流時，脂蛋白-X 是膽汁淤積中導致高脂血症的主要原因。有別於低密度脂蛋白（俗稱「壞膽固醇」），脂蛋白-X 不含載脂蛋白 B，而載脂蛋白 B 是低密度脂蛋白的主要成分，也是動脈粥樣硬化斑塊發病機制中最重要的因素之一，因此脂蛋白-X 不會導致動脈粥樣硬化。此外，基於相同原因，由於載脂蛋白 B 是肝臟低密度脂蛋白受體最重要的配體，而脂蛋白-X 缺少載脂蛋白 B，因此不能內化到肝細胞中。由於脂蛋白-X 高膽固醇血症不是由於肝細胞過度產生所導致，因此使用他汀類藥物等藥物來減少膽固醇合成是無效的，也沒有必要。

醫療團隊報告了兩個本地病例，患者均為曾接受造血幹細胞移植的中國籍兒童。醫療團隊盼負責移植手術的醫生及內分泌科專科醫生日後能多加留意，造血幹細胞移植後肝移植物抗宿主病患者或會患上由脂蛋白-X 導致的嚴重高膽固醇血症，故應小心使用他汀類藥物，以免藥物未能發揮功效。

詳細內容可參閱原文《造血幹細胞移植後肝移植物抗宿主病中國籍兒童患者的脂蛋白-X 高血脂症：兩個病例報告》。

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