

Changing Nomenclature for PBC from “Primary Biliary Cirrhosis” to “Primary Biliary Cholangitis”

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Primary biliary cirrhosis (PBC) is a chronic cholestatic liver disease, frequently developing in middle-aged women [1]. In patients with PBC, small bile ducts in the liver are believed to be damaged via autoimmune reactions [2]. Patients were diagnosed as having PBC when 2 or more among the following 3 features are observed; elevated cholestatic enzymes, presence of anti-mitochondrial autoantibodies (AMA) and histological findings as chronic non-suppurative destructive cholangitis (CNSDC) in

particular [3]. When the disease entity of PBC was established more than 50 years ago [4], advanced liver diseases showing histological findings of cirrhosis were found in the most of patients with PBC. Consequently, a nomenclature for primary biliary “cirrhosis” was reasonably accepted by all hepatologists and gastroenterologists world-widely.

Introduction of AMA as a tool for screening examination, however, enabled physicians to diagnose patients with chronic cholestatic diseases as having PBC during the earlier stages before developing cirrhosis [5]. Moreover, spread use of ursodeoxycholic acid (UDCA) as a first-line therapeutic agent, contributed to inhibit the progression of liver diseases from the non-cirrhotic stage to the cirrhotic stage in a large part of patients with PBC [6]. In Japan, about 70–80 % of patients with PBC were shown to be diagnosed as having “asymptomatic PBC” in the recent years [3]. Thus, the nomenclature of primary biliary “cirrhosis” is regarded as a misnomer both from clinical and histological aspects. As a result, the patients inevitably misunderstand the stage of their diseases and at the same time have to bear the substantial burden in their social activities.

During 2014 and 2015, the governing boards of both the European Association of the Study for the Liver (EASL) and American Association of the Study for the Liver (AASLD) have approved the change of nomenclature for PBC from “primary biliary cirrhosis” to “primary biliary cholangitis”, and published the statements in the official journals [7, 8], followed by publishing identical articles in several major journals in the fields of gastroenterology and Hepatology [9–11]. In contrast, in Japan, the Intractable Hepato-Biliary Diseases Study Group supported by Health Labor Science Research Grants from Research on Measures for Intractable Diseases performed the questionnaire survey regarding the nomenclature for PBC on 18

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experts for clinical research of PBC in 2014. Although all the experts agreed to revise the nomenclature for PBC, unanimous consent was not obtained. Of the 18 Japanese experts, 11 supported the nomenclature of “cholangitis”, while 5 and 2 experts proposed “cholangiopathy” and the other nomenclatures, respectively. “Biliary” and “cholangitis” may sound as redundant repetition, and “cholangitis” does not reflect pathological features observed in the liver of patients with PBC exactly. Considering consistency with the nomenclature approved by EASL and AASLD, the Study Group decided to adopt the nomenclature of “primary biliary cholangitis” to keep a well-known abbreviation “PBC”. However, an alternative nomenclature for PBC, which more accurately represents the disease entity, should be established in the future.

In December 2015, the Intractable Hepato-Biliary Diseases Study Group of Japan addressed the proposal regarding revision of nomenclature for PBC to the governing boards of the Japanese Society of Gastroenterology (JSGE) and the Japan Society of Hepatology (JSH), and the nomenclature of “primary biliary cholangitis (PBC)” was approved in January 2015 and March 2015 by JSH and JSGE, respectively. Both societies proposed the novel nomenclature jointly to the Japanese Association of Medical Sciences (JAMS) in April 2014, and the nomenclature of “primary biliary cholangitis”, will be officially used by the Ministry of Health, Labor and Welfare of Japan following discussion by the committee for regulation of medical terms organized in JAMS.

We sincerely call on all members of the Japanese Society of Gastroenterology and the Japan Society of Hepatology to use from this moment on the name “primary biliary cholangitis”, instead of primary biliary “cirrhosis”, for the disease known by its abbreviation PBC.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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