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Experts consensus on Chinese nomenclature of Budd-Chiari syndrome

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ABSTRACT

In China, Budd-Chiari syndrome has been transliterated into six names according to the pronunciation of the letters. To standardize and unify the Chinese names of the disease, multi-disciplinary experts suggest translating Budd-Chiari syndrome into hepatic vein inferior venal cava obstruction syndrome as its Chinese name after reaching a consensus through discussion.

1. The original generalized Budd-Chiari syndrome

In 1845, Budd reported autopsy findings of extensive hepatic vein occlusion and thrombosis, which had led to hepatosplenomatosis and portal hypertension; Chiari reported similar findings in 1899. Later literature stated that oral contraceptive pills, Behcet's disease, and gene mutations could also cause this syndrome, commemorate the discovery by Budd and Chiari, the disease is called Budd-Chiari syndrome.[1,2] It known as the primary definition of Budd-Chiari syndrome.In1968, Nakamura et al. found that inferior vena cava obstruction above the hepatic vein opening could also produce posthepatic portal hypertension. Therefore, the obstruction at any point from the efferent acinar vein to the end of the inferior vena cava causes Budd-Chiari syndrome. This definition is still in use. The generalized Budd-Chiari syndrome is a series of clinical manifestations caused by obstruction of venous return between the hepatic sinus vein and the posterior hepatic segment of the inferior vena cava.[3–6]

The expansion of the definition from original to generalized Budd-Chiari syndrome was possible because understanding of the condition has continuously improved, mainly because clinical diagnosis technology has progressed from histological techniques at autopsy and liver biopsy to non-invasive imaging methods. Furthermore, the clinical treatment of Budd-Chiari syndrome has transformed from ineffective conservative management and traumatic surgery to effective intervention. According to both domestic and foreign literature from the past 30 years, China has the largest number of patients with Budd-Chiari syndrome, with more than 20,000 cases, and inferior vena cava obstruction is principle characteristic of the condition in East Asian patients. Thanks to interventional therapy and technological innovations, acutely life-threatening Budd-Chiari syndrome is no longer an incurable disease. Moreover, Chinese imaging diagnosis and interventional treatment of Budd-Chiari syndrome has attained an international leading position,[7] As such a standardized Chinese name and abbreviation of Budd-Chiari syndrome must be devised.

2. Epidemiology, etiology, and pathology of Budd-Chiari syndrome

Domestic literature has shown that the middle and lower reaches of Huang-Huai region have the highest incidence of Budd-Chiari syndrome in China.[8–10] Zhonghao et al. conducted a census of 680,000 people to identify the incidence of Budd-Chiari syndrome in Dongping County. A 1986 study in Shandong Province used epidemiology, etiology, pathology, clinical classification, diagnosis, and treatment data to determine that the incidence in the region was 4.4/100,000,[11] Zu Maoheng et al. treated 952 patients admitted to three hospitals in Xuzhou city over the past 30 years. By the end of 2019, The Xuzhou population was 9.06 million and the clinical epidemiological incidence was 10.5/100,000. By the end of 2019, the resident population of Xiao County, Anhui Province was 789,000, among whom 115 patients received interventional treatment for Budd-Chiari syndrome in the Affiliated Hospital of Xuzhou Medical University. Therefore, the clinical epidemiological incidence was 14.57/100,000. This is consistent with the report by Zhang Wei et al. who found more than 20,000 cases of hepatic vein and inferior vena cava

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obstruction in the Chinese literature, 80% of which were from Jiangsu, Shandong, Henan, and Anhui provinces,[12] Many cases in China have shown membranes at the opening of the hepatic vein and near the right atrium of the inferior vena cava.These membranes lead to segmental occlusion that can involve the renal vein level only or the entire inferior vena cava. Membrane formation is the characteristic pathological change in Budd-Chiari syndrome in China. However, the etiology and pathogenesis of membrane formation and evolution remain unclear.[13]

3. Budd-Chiari syndrome translation into Chinese

Before 1980 due to historical limitations and a lack of diagnostic tools, Budd-Chiari syndrome was poorly recognized in China. The first report in 1963 and 1981 described Budd-Chiari syndrome in only 41 cases across China.[14] As such it was considered an extremely rare disease, and most doctors in China knew too little about Budd-Chiari syndrome, so the first-opinion misdiagnosis rate was as high as 44%–97.5%.[15,16] Because so few cases were reported, the Chinese translation of Budd-Chiari syndrome failed to attract people's attention.

In 1988, Li Yanhao et al. reported the X-ray diagnosis and treatment of 140 cases of Budd-Chiari syndrome,[17] and the number of domestic reports involving large groups of cases increased every year subsequently,[18–21] as did the use of the Chinese name of Budd-Chiari syndrome. The disease name has been translated phonetically into Chinese as *bu-jia* syndrome,[22] *bu-ka* syndrome,[23] *bai-cha* syndrome,[24] *bad-jiliya* syndrome,[25] *bad-jiyali* syndrome,[26]*and bad-qiari* syndrome,[27] Of course, it ti inappropriate for a disease to have more than one name. Moreover, using transliterations of disease names can be problematic for the following reasons, (1) Different accents in different parts of China lead to different transliterations, this is not conducive to academic exchange or big data application. (2) In recent years, most Chinese literature has called the condition *bu-jia* syndrome, while the name of the disease is *bad-jiyali* syndrome in China's medical record management, such a big difference is not conducive to clinical work. (3) Having different Chinese translations is not conducive to medical record management or medical insurance management. In the medical record management system, the disease is called *bad-jiyali* syndrome, and the code is 182.0. This confuses people as they do not know the meaning. In a Chinese encyclopedia about medical syndrome, there are more manes as Budd - and Budd-Chiari syndrome disease, other name as Chiari syndrome, hepatic venous occlusive thrombosis, inferior vena cava obstruction syndrome, Roki-Tansky syndrome, hepatic vein occlusion syndrome, hepatic vein thrombosis syndrome, hepatic vein occlusion disease, inferior vena cava occlusion with hepatic vein obstruction syndrome, hepatic vein reflux obstacle syndrome of 11 different name.[28] (4) The literal meaning of the Chinese translated names has nothing to do with the disease itself; that is, the Chinese translations do not evoke the characteristics of hepatic vein and inferior vena cava obstruction, nor do they reflect the differences in the disease between East Asia and the west, this is not conducive to clinical application.

Foreign definitions of Budd-Chiari syndrome focus on hepatic vein obstruction,[29] while Chinese definitions refer to obstruction on the inferior vena cava. The existing translations also fail to reflect the meaning and pathological characteristics of Budd-Chiari syndrome.

India and Nepal also have a high incidence of Budd-Chiari syndrome,and researchers there also report blocking of the vena cava primarily as in China. They refer to Budd-Chiari syndrome as hepatic vena cava syndrome.[30] In 2010 the Chinese academic Zhonghao asserted that "Budd-Chiari syndrome is a posthepatic portal hypertension caused by obstructive lesion above the opening of the hepatic vein and/or inferior vena cava that are often accompanied by inferior vena cava syndrome. As such it is more accurate to call it hepatic vena cava syndrome."[7]

4. New Chinese names and definitions of Budd-Chiari syndrome

Standardization and normalization of disease diagnostic names are crucial to health informatization[31] and internet medical care which is in line with the national medical insurance policy of "spending money on patients who have special needs and ensuring that medical fees are paid for more people, namely: those with single disease or multiple diagnoses within related groups."[32]

According to the provisions of disease nomenclature, the naming of diseases includes four elements: anatomical location, pathogenic factors, pathological changes, and clinical manifestations. In more than 20,000 clinical cases in China, Budd-Chiari syndrome has involved obstruction of the hepatic vein and/or inferior vena cava, so we propose using the Chinese name "hepatic vein inferior vena cava obstruction syndrome," which is further characterized by consequent posthepatic portal hypertension and/or inferior vena cava hypertension. The English translation also calls Budd-Chiari syndrome, must be emphasized, although it must also be clarified that the intent is not to rename Budd-Chiari syndrome or modify the original definition, but rather to reform the Chinese name of the disease with the provisions of this standard, to consolidate the different Chinese name that result from different accents across the country, and thus to devise a single Chinese name and definition. The proposed new Chinese name emphasizes hepatic vein and/or inferior vena cava obstruction at the hepatic vein opening including anatomical and/or hemodynamic obstruction. As such, it conforms to the relevant provisions of disease naming, as well as objectively and clearly reflecting the disease characteristics and the necessity of clinical treatment.

Use of the term "Budd-Chiari syndrome" in oral and written communication does not conflict with the Chinese name of this article.

Hepatic vein and/or inferior vena cava obstruction caused by tumor invasion of the hepatic vein or tumor thrombus can be called secondary hepatic vein and inferior vena cava obstruction syndrome.[33,34]

The pathological mechanism of hepatic vein occlusion caused by pyrrolidine and other drugs is clear. Treatment methods include anticoagulants, liver protection, and transjugular intrahepatic portosystemic shunt. The lesion has been included in the category of drug-induced liver injury, and is called hepatic sinus obstruction syndrome. It is no longer considered a subtype of hepatic vein inferior vena cava obstruction syndrome and is attributed to posthepatic portal hypertension.[35,36]

Inferior vena cava occlusion below the renal vein level is still called inferioe vena cava occlusion syndrome because it does not affect hepatic venous blood return or cause posthepatic portal hypertension.

The above consensus was discussed and confirmed by Chinese experts in pathology, imaging diagnostics, vascular surgery, interventional radiology, and hepatobiliary surgery.

Experts in the discussion:

Ce Bian (General hospital of PLA Rocket Force), Jianmin Cao (Eastern theater hospital of Chinese PLA), Yongde Cheng (Interventional radiology journal editorial office), Jinguo Cui (Chinese PLA 980 hospital), Xiaowei Dang (The first affiliated hospital of Zhengzhou university), Jiahong Dong (Tsinghua Changgung hospital), Yong Gao (The affiliated hospital Bengbu medical college), Jianping Gu (Nanjing first hospital) Yuming Gu (The affiliated hospital of Xuzhou medical university), Chenghao Guo (Shandong university), Xinwei Han (The first affiliated hospital of Zhengzhou university), Linsun Li (The first affiliated hospital Nanjing medical university), Yanhao Li (Southern hospital), Zhen Li (The first affiliated hospital of Zhengzhou university), Zuoqin Liu (Shandong province people's hospital), Wei Mu (The first Affiliated Hospital of Army Medical University), Caifang Ni (The first affiliated hospital of Suzhou university), Zhonggao Wang (General Hospital of PLA Rocket Force), Maoqiang Wang (The general hospital of Chinese PLA), Hua Xiang (Hunan province people's hospital), Hao Xu (The affiliated hospital of Xuzhou medical university), Kai Xu (The affiliated hospital of Xuzhou medical university), Ke Xu (The first affiliated hospital of China medical university), Zhiping Yan (Zhongshan hospital), Lin Zhang (Tsinghua Changgung hospital), Qingqiao Zhang (The affiliated hospital of Xuzhou medical university), Xiaoming Zhang (Beijing people's hospital), Maoheng Zu (The affiliated hospital of Xuzhou medical university).

Declaration of competing interest

The authors declare that they have no conflicts of interests to this work. We declare that we do not have any commercial or associative interest that represents a conflict of interest in connection with the work submitted.

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