Compensation by collateral circulation determines invasive therapeutic indications for patients with Budd-Chiari syndrome

We read with interest the comprehensive review article by Rössle and Gerbes that details the management of ascites in patients with liver cirrhosis and concludes that the transjugular intrahepatic portosystemic shunt (TIPS) could manage refractory ascites more effectively than large-volume paracentesis. However, there is an important issue regarding the management of ascites, which is caused by Budd-Chiari syndrome (B-CS), that the authors failed to address.

In patients with chronic course, the formation of intra and extrahepatic collaterals leads to improvement of liver function and may silence this disease and make it asymptomatic. Thus, collateral circulation may be of great clinical importance in guiding the treatment of B-CS. Our clinical study demonstrated that TIPS was not necessary in chronic cases of B-CS.

Data were available for a total of 174 B-CS patients (39% women, mean age 37 years) treated between 1998 and 2011. The imaging analysis demonstrated various levels of collateral circulation in these patients. We classified B-CS patients into six pathophysiological subtypes, according to their haemodynamic changes and compensational situation caused by collateral circulation, and different treatment strategies were also outlined (table 1). Among these patients, 12 did not undergo any special treatment (6.9%), 38 of the patients underwent radiological intervention (RI) and 21 patients underwent surgical procedures (SPs). Staged treatments were adopted in 103 cases. Sixty-one of the 103 patients underwent RI and 13 underwent SPs due to inferior vena cava hypertension (IVCHT); one patient was treated for portal hypertension (PHT). Additionally, 29 patients underwent second-stage treatment: RI and SPs were performed in 25 cases, and four patients underwent SPs alone. The incidences of morbidity and mortality of patients undergoing RI were much lower than those observed for patients undergoing SPs (4.4% vs 25.6%, p<0.01).

During follow-up, four patients received second-stage treatment, and 12 cases underwent IVC/middle hepatic vein recanalisation. In addition, two patients died of hepatocellular carcinoma, and three patients died of graft obstruction. The liver biopsy results from 47 patients, three to six years postoperatively, demonstrated significant improvement in their fibrosis grade. When compared with patients treated with TIPS, our new strategy showed great advantages in terms of long-term survival.

Our findings also support the concept of stage management for B-CS with IVC and main hepatic vein obstruction. In the past, direct decompression of PHT was the treatment priority. Mesoatrial, mesojugular and splenoatrial SPs were performed and achieved higher rates of perioperative mortality, morbidity and poor long-term survival. In our study, the first-stage treatment priority was IVCHT. After recanalisation of the IVC, the necessity of second-stage treatment for PHT was
assessed according to changes in clinical manifestations and biochemical tests. In these patients with symptoms and signs of IVCHT and PHT, collaterals were often identified in patients who had undergone first stage treatment than in those who had second stage treatment, which led to complete compensation for PHT after the recanalisation of IVC. The stage management strategy could restore a physiological haemodynamic state, which TIPS could not.

To our knowledge, this is the first report on the role of collateral circulation, which was pivotal in the management strategy. Using the compensational status caused by these collaterals, a pathophysiological classification and a different management strategy for B-CS was generated and achieved satisfactory results. These results demonstrated that the compensated status, by collateral circulation, determined the invasive therapeutic indication for patients with B-CS. In addition, this study also highlighted the advantages of regular RI in the management of this disease.

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Contributors YS and XM: conception and design, collection and assembly of data, data analysis and interpretation, manuscript writing, final approval of the manuscript; LF: data interpretation and manuscript revision; SG and ZW: collection and assembly of data, data analysis and interpretation.

Funding This study was supported by a grant from the National Natural Science Foundation of China (81100304) and the Projects of the Ministry of Public Health (No. 201002015).

Competing interests None.

Ethics approval Ethics approval was provided by the Ethics Committee of Zhengzhou University.

Provenance and peer review Not commissioned; externally peer reviewed.

Accepted 3 May 2012
Published Online First 11 June 2012
doi:10.1136/gutjnl-2012-302471

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