

# Pathogenetic Classification of Portal Hypertension \*

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ALTHOUGH SOME investigators advanced a *forward-pressure* theory for certain types of portal hypertension,<sup>6, 12, 21, 23, 25, 26, 27, 28, 35</sup> pressure elevation within the portal system is commonly regarded as being caused by an increased resistance. This concept is corroborated by measurements of hepatic blood flow which apparently is normal or below normal in this condition.<sup>10, 32</sup>

Whipple in 1945 classified diseases with portal hypertension according to the site of increased resistance into intra- and extrahepatic block.<sup>37</sup> This classification was later supplemented by differentiating extrahepatic block into the pre- and posthepatic type.<sup>17, 36</sup>

Certain drawbacks of this simple classification have prompted Sherlock<sup>33</sup> and Imanaga and co-workers<sup>15</sup> to subdivide prehepatic presinusoidal, intrahepatic presinusoidal, intrahepatic postsinusoidal and extrahepatic postsinusoidal block. The main reason for this new classification was that cirrhosis of the liver in some instances may give rise to presinusoidal block<sup>3</sup> and in other cases leads to postsinusoidal block.<sup>7, 29</sup> Other reasons are that schistosomiasis, a hepatic disease, causes presinusoidal block and probably the same is true of congenital liver fibrosis, Hodgkin's disease and sarcoïdosis.<sup>18, 32, 33</sup>

There is a fluent transition between these groups, especially in cirrhosis of the liver. Moreover, for the surgeon it is of no consequence whether there is pre- or postsinusoidal block, for shunt operation is his objective in any case. Only if the vena cava

is affected in hepatic vein block, pressure in this vessel rises and shunt operations become useless. It is evident (Table 1) that vena caval involvement in cases with hepatic vein thrombosis is more frequent than isolated hepatic vein thrombosis. Combination of thromboses in different vessels of the portal system and combination of portal vessel thrombosis with thromboses in the systemic circulation is surprisingly frequent (Table 1).

TABLE 1. *Relative Frequency of Thrombosis in the Portal System*

	No. cases	%
Isolated portal vein thrombosis	107	32.7
Portal and splenic vein thromboses combined with mesenteric vein thrombosis	63	19.3
Portal and splenic vein thromboses	46	14.1
Portal system thromboses with thromboses in the systemic circulation	20	6.1
Isolated hepatic vein thrombosis	28	8.6
Hepatic vein thrombosis combined with thromboses in other vessels	59	18.0
with inf. V. cava	15	
with inf. V. cava and other thrombosis	20	
with thromboses in systemic circulation	24	
Isolated splenic vein thrombosis	4	1.2
Total	327	

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Various diseases such as polycythemia, rheumatic phlebitis, luetic phlebitis, leukemia and migratory phlebitis may involve prehepatic as well as posthepatic vessels. In the light of these facts, therefore, classification of portal hypertension as to the site of increased resistance appears to be of doubtful value.

Advances in diagnosis since Whipple's paper, especially routine splenoportography, needle biopsy of the liver and peritoneoscopy, have made possible a more precise differentiation of diseases with portal hypertension. Table 2 lists and classifies diseases which may be accompanied by pressure rise in the portal system.

With this pathogenetic classification portal hypertension is regarded as a symptom of various disease processes, all having individual clinically important characteristics. Characterization of the process is best accomplished from the diagnosis of cirrhosis of the liver, splenic or portal vein thrombosis, hepatic vein thrombosis, etc. and the designation pre-, intra- or posthepatic are superfluous.

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TABLE 2. *Diseases in Which Portal Hypertension\* Occurs*

DISEASES OF THE LIVER	
Congenital	
Hypoplasia <sup>4</sup>	
Polycystic disease <sup>22</sup>	
Congenital fibrosis <sup>18</sup>	
Hepatitis <sup>11</sup>	
Cirrhosis <sup>12</sup>	
Parasitic	
Schistosomiasis <sup>2</sup>	
Echinococcosis <sup>5</sup>	
Tumors of the liver	
primary <sup>30</sup>	
metastatic <sup>31</sup>	
Gummata of the liver <sup>13</sup>	
Systemic diseases with liver involvement	
Sarcoidosis <sup>8</sup>	
Lymphogranulomatosis <sup>19</sup>	
Hematological disorders <sup>32</sup>	
Lipidoses <sup>16</sup>	
Reticuloendothelioses <sup>32</sup>	
VENOUS OCCLUSION IN THE PORTAL SYSTEM	
Congenital <sup>20</sup>	
Thromboses <sup>1</sup>	
Venous compression <sup>14</sup>	
HEART DISEASES WITH PORTAL HYPERTENSION <sup>9</sup>	
ANEURYSMS OF SPLENIC AND HEPATIC ARTERY WITH PORTAL HYPERTENSION <sup>24</sup>	
ARTERIOPORTAL FISTULAS AND PORTAL HYPERTENSION <sup>34</sup>	

\* A more complete list of references to diseases with portal hypertension may be obtained from the author.

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