VASCERN HHT

Management of Hepatic arteriovenous malformations, Dubrovnik, Croatia, June 2017

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The prevalence of hepatic malformations (HVMs) is particularly high in patients with a pathogenic *ACVRL1* mutation; whereas only 8% have symptomatic HVMs in cross sectional studies. Longitudinal series have shown an incidence of 3.6% per year of complications and a 1% mortality per year due to liver VMs in HHT.

Only patients with symptomatic HVMs should be treated.

FIRST LINE TREATMENT

The first line treatment consists of

(i) treatment of high output heart failure should be similar to what is done for heart failure outside HHT, with special emphasis on escalating diuretics dosages;
(ii) treatment of portal hypertension is equal to the treatment of biliary cirrhosis, and
(iii) antibiotics in case of cholangitis.

The results of these first line intensive treatments are to be judged within 6-12 months.

LIVER TRANSPLANTATION

If complicated liver VMs are refractory to first line treatment, liver transplantation is to be evaluated, whereas trans-arterial embolization is generally not an option due to its palliative effect and inherent risk of liver necrosis.

Main indications for liver transplantation in the cases so far reported in literature have been high output heart failure, complicated portal hypertension and biliary ischemia.

Literature data show that liver transplantation for complicated liver VMs has good outcome resulting in hemodynamic and clinical normalization and improved quality of life.

Available data suggest that liver transplantation should be timely proposed before pulmonary resistances become fixed, and taking into account that complicated liver VMs in HHT represent a MELD-exception for transplantation.

Boillot addressed surgical particularities in liver transplantations for HHT patients. Amongst other, the recipient hepatic artery should be removed and the donor artery should anastomosed to the origin of the recipient hepatic artery on the coeliac trunk, in order to prevent hepatic artery aneurysms, rupture and/or thrombosis.

BEVACIZUMAB

The use of Bevacizumab before liver transplantation does not change the surgical procedure but the interval between Bevacizumab and transplantation should be preferably 2-3 months.

CONCLUSIONS

On the basis of available literature data, it was concluded that

i. Liver transplantation might be indicated in patients under 65 years old;
ii. In patients younger than 65 years not fit for transplantation, Bevacizumab could be tried and used as a "bridge-to- transplantation" if patients respond;
iii. In patients older than 65 Bevacizumab can be proposed, with following maintenance dosages if patient respond [1].

The decision to proceed to either liver transplantation or bevacizumab has to be discussed with the patient, by clearly weighing the risk benefit balance, on a case-by-case basis, and in the context of a center with specific HHT expertise.

REFERENCES

[1] European Association for the Study of the Liver. Electronic address: easloffice@easloffice.eu (2016) EASL Clinical Practice Guidelines: Vascular diseases of the liver. J Hepatol 64:179-202

Published within

THE EXECUTIVE SUMMARY OF THE 12TH HHT INTERNATIONAL SCIENTIFIC CONFERENCE
https://www.ncbi.nlm.nih.gov/pubmed/29147802

Buscarini and Dupuis-Girod 2017
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