Journal Watch

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- **ESPGHAN Position Paper on Management and Follow-up of Children and Adolescents with Celiac Disease (CD).**

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The aim of this position paper was to gather the current evidence and to offer recommendations for follow-up and management of children and adolescents with CD. On the basis of 164 publications and expert opinions, 37 recommendations were formulated. The salient features are as follows:

- After the diagnosis is established a regular follow-up by an expert physician or dietician should be done. The first follow-up should be done after 3–6 months of diagnosis, later every 6 months until normalization of IgA-tissue transglutaminase (IgA-TGA) levels, and every 1–2 yearly thereafter.
- During follow-up signs and symptoms should be evaluated along with anthropometric parameters. IgA-TGA should be done as a marker for mucosal healing. Screening for thyroid disease can be done with thyroid stimulating hormone (TSH). Other blood investigations should be done if required on the basis of symptoms. Health-related quality of life should be measured in all children.
- Adherence to a gluten-free diet (GFD) should be evaluated on the basis of symptoms, dietary history, and TGA levels. The role of gluten immunogenic peptides is not yet established to assess adherence.
- If significant catch-up growth is not attained within 1 year of GFD then an alternative diagnosis should be sought.
- Supplementation should be given to children with iron , folate or Vitamin B12, and improvement should be documented.
- Persistent high IgA-TGA should be evaluated by dietary compliance, retesting, and evaluation for other enteropathies.
- Re-biopsy should be considered only in cases of doubts in diagnosis or suspicion of another disease.
- Gluten challenge should be done in children with an uncertain diagnosis but it should be avoided during the period of accelerated growth. Ingestion of 10–15 gm/day of gluten for 3–6 months is expected to induce mucosal abnormality in the majority of children with CD. The optimum amount and time are not yet clear. To avoid unnecessary exposure IgA-TGA should be measured 1 month after the challenge and then every 3 months for 1 year. In case of symptoms, mucosal biopsy should be performed else the child should be subjected to a normal diet after 1 year of challenge.

- **EASL Clinical Practice Guidelines on Prevention and Management of Bleeding and Thrombosis in Patients with Cirrhosis.**

(J Hepatol. 2022 May; 76(5):1151–1184. DOI: 10.1016/j.jhep.2021.09.003. Epub 2022 Mar 15. PMID: 353,00861.)

This guideline was formulated to clear the confusion regarding some debated topics such as hemostasis in liver disease, correction of thrombocytopenia for invasive procedures, and thromboprophylaxis. Following are the recommendations:

- Patients with cirrhosis and deranged laboratory parameters ([international normalised ratio (INR), activated partial thromboplastin clotting time (APTT), platelet count, fibrinogen]) can present with clinically relevant spontaneous bleeding as intracranial hemorrhage, muscular or orbital hematomas, spontaneous hemoperitoneum or arterial bleeding, etc. Administration of Vitamin K has not been shown to correct the prolongation of INR in cirrhotic patients, however, it does transiently improve INR in cholestatic liver disease. Also, currently, there is no evidence that fresh frozen plasma (FFP) or platelet transfusion prevents spontaneous bleeding.
- Alteration of coagulation profile (INR, platelet count, fibrinogen) is often seen in cirrhotics, the main reasons being hypersplenism and decreased hepatic production of thrombopoietin. So far, no association has been proven between altered coagulation profile and procedure-related bleeding in cirrhotic patients. These tests can define the severity of the disease and the hemostatic status of the patient. So, laboratory evaluation of the hemostatic profile is not recommended to predict post-procedural bleeding. Also, correction of prolonged INR with FFP or correction of fibrinogen deficiency in order to decrease post-procedural bleeding is not recommended. In case of thrombocytopenia in patients undergoing high-risk procedures, if the platelet count is between 20×109 /L and 50×109 /L infusion of platelet concentrates or thrombopoietin receptor agonists should not be routinely performed and should be individualized.
- Clinical studies have shown that the risk of developing deep vein thrombosis/pulmonary edema (DVT/PE) is as high as the general population. Risk can be assessed using clinical scores as Padua prediction score or IMPROVE score (validated in the adult population). The use of laboratory tests to predict DVT or PE is not recommended.

- **EASL Clinical Practice Guidelines on The Management of Hepatic Encephalopathy (HE).**

(J Hepatol. 2022 Sep; 77(3):807–824. DOI: 10.1016/j.jhep.2022.06.001. Epub 2022 Jun 17. PMID: 357,24930.)

This practice guideline was put together after evaluating 726 publications by experts. Important noteworthy points are as follows:

- Classification: HE is classified on the basis of etiology: type I (due to acute liver failure), type II (portosystemic shunt without significant liver disease), and type III (cirrhosis with or without portosystemic shunt); on the basis of severity: covert (minor or no signs/symptoms but abnormalities on neuropsychological and/or neurophysiological tests) and overt (grades II or over according to the West Haven criteria2); On the basis of time course: episodic, recurrent (more than one episode over a period of 6 months), or persistent (no return to normal/baseline neuropsychiatric performance in between episodes).
- Grading: For grading and staging in clinical practice West Haven criteria should be used at least from grade II upwards. For patients with impaired consciousness and patients in the intensive care unit, the Glasgow coma scale should be applied.
- Plasma ammonia levels: In all patients with encephalopathy and liver disease plasma ammonia should be measured. A normal value has a negative predictive value and it should indicate an alternative diagnosis for the encephalopathy. However, hyper-ammonia may be seen without HE or any liver disease. Ammonia levels correlate with the severity of HE but it should not be used to monitor therapy. High levels are also associated with decreased transplant-free survival.
- Brain imaging: There is no specific feature on imaging of the brain in HE patients, so it should not be used for diagnosis of HE, but it can help in case of suspicion of other brain lesions and can provide information such as cerebral edema or atrophy.
- Liver support system: In patients with acute liver failure high-volume plasma exchange has been shown to improve the grade of HE and confers a survival benefit, but this is not demonstrated in patients with cirrhosis.
- Prevention: Decompensation of cirrhosis is strongly associated with mortality in HE patients. Management of decompensation of the underlying liver disease as acute variceal bleed improves prognosis. Also, prevention or management of the precipitating factors is associated with recovery from HE in 90% of patients.
- Secondary prophylaxis: In patients who have recovered from HE recurrence can be prevented by lactulose or rifaximin.
- Supplementation: Patients with cirrhosis are prone to water-soluble vitamin deficiency. Oral vitamin supplementation is justified in cirrhotics. There is no role of zinc supplementation for improvement in mental status.
- Dietary change: In patients with recurrent HE and intolerance to animal protein, replacement with vegetable or dairy protein can be tried along with monitoring.
- Liver transplantation: Patients with complicated or recurrent episodes of HE should be considered for liver transplantation.

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