P1354 A SYSTEMATIC LITERATURE REVIEW (SLR) OF THE MANIFESTATIONS OF VENO-OCCLUSIVE DISEASE/SINUSOIDAL OBSTRUCTION SYNDROME (VOD/SOS) AFTER HEMATOPOIETIC CELL TRANSPLANTATION (HCT) IN ADULTS VERSUS CHILDREN

Topic: 22. Stem cell transplantation - Clinical

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Background: VOD/SOS is a potentially fatal complication of HCT. Its diagnosis can be challenging due to the many patient- and transplant-related VOD/SOS risk factors, its sometimes slow, progressive onset, and the dynamic presentation of signs and symptoms, some of which may differ by age.

Aims: This SLR evaluated differences in the clinical manifestations of VOD/SOS post-HCT in adults compared to children.

Methods: Medline and Embase were searched to 4 March 2021 for studies of VOD/SOS post-HCT and were supplemented with guidelines on VOD/SOS diagnosis and management. English language reports of observational and database studies were included if they studied adults or children with any HCT-related disease, therapies aimed at preventing/treating VOD/SOS, or HCT-associated VOD/SOS outcomes. Publications were evaluated based on inclusion of the 5 cardinal features related to VOD/SOS diagnostic criteria (i.e., ascites, hepatomegaly, bilirubin ≥2 mg/dL, weight gain of ≥5%, and right upper quadrant [RUQ] pain) and changes in diagnostic features over time. In cohort studies, an unweighted mean was calculated for the proportion of patients with each VOD/SOS feature; for case studies, the number of case reports in which the feature was present was divided by the total number of cases in which any features were reported.

Results: Overall, 204 publications were included in the SLR. Weight gain of ≥5% and hepatomegaly were more common at diagnosis in children compared to adults, whereas the presence of ascites or RUQ pain was similar between age groups (Figure). While only 40% of cases had all 5 cardinal features of VOD/SOS, age did not determine the likelihood of missing any single feature. When weight gain was present, it appeared as the first manifestation more often in adults than children (54% vs 22%, respectively); it was more likely to appear second in children, suggesting VOD/SOS-related weight gain may be less easily detected or develops later in children. Similarly, hepatomegaly appeared first in >90% of adults compared to <50% of children. While RUQ pain was more likely to be the second or third feature in children, it appeared first in around 85% of adults compared to 50% of children, suggesting it presents in children in relation to other symptoms or is more difficult to detect. Ascites was absent in 22.7% of all cases, making it the feature most frequently missing; however, when it was present, it appeared as the first manifestation more frequently in children than adults (40% and 6%, respectively). There was no difference between adults and children on the day post-HCT that the first VOD/SOS features were detected.

Hyperbilirubinemia was more common at diagnosis in adults than children (Figure). Importantly, hyperbilirubinemia was absent in 18.2% of children and 6.7% of adults, and analysis of individual VOD/SOS features showed younger age was associated with an absence of hyperbilirubinemia compared to other features. The averaged time series data for bilirubin levels appeared to peak earlier and to have a higher peak for adults than children, suggesting more rapid onset in adults.

Image:
Summary/Conclusion: Presentation of VOD/SOS symptoms post-HCT, as identified and reported in publications of patients diagnosed with VOD/SOS, varied between adults and children. Notably, VOD/SOS without elevated bilirubin may be more common at diagnosis in children compared to adults. VOD/SOS features are not reported in a distinct sequence, requiring a high index of suspicion for VOD/SOS and continuous vigilance by those involved in patient monitoring post-HCT.