

Pediatric chronic graft-versus-host disease

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Cuvelier GDE, Nemecek ER, Wahlstrom JT, Kitko CL, Lewis VA, Schechter T, Jacobsohn DA, Harris AC, Pulsipher MA, Bittencourt H, Choi SW, Caywood EH, Kasow KA, Bhatia M, Oshrine BR, Flower A, Chaudhury S, Coulter D, Cheung JH, Joyce M, Savaşan S, Pawlowska AB, Megason GC, Mitchell D, Cheerva AC, Lawitschka A, West LJ, Pan B, Al Hamarneh YN, Halevy A, Schultz KR. Benefits and challenges with diagnosing chronic and late acute GVHD in children using the NIH consensus criteria. *Blood*. 2019;134(3):304-316.

1. Your patient is a 12-year-old boy thought to have chronic graft-versus-host disease (cGVHD) after allogeneic hematopoietic stem cell transplantation. According to the prospective multi-institution study by Cuvelier and colleagues, which of the following statements about use of the National Institutes of Health Consensus Conference (NIH-CC) criteria to diagnose pediatric cGVHD is correct?

- NIH-CC criteria are unsuitable to diagnose pediatric cGVHD
- NIH-CC criteria confirmed 41% of 39 reported cGVHD cases but not 23 (59%) cases (10 reclassified as late acute graft-versus-host disease [L-aGVHD]; 5 unrelated to graft-versus-host disease; 8 deemed probably/likely cGVHD but not meeting NIH-CC criteria)
- The most common reason for reclassification of submitted cGVHD cases was negative biopsy results
- Patients thought to have cGVHD but not meeting NIH-CC criteria likely had mild disease with extremely late presentation

2. According to the prospective, multi-institution study by Cuvelier and colleagues, which of the following statements about the frequency of cGVHD, L-aGVHD, and pulmonary cGVHD in children determined by using the NIH-CC criteria is correct?

- After study committee review and application of NIH-CC criteria, only 16% of reported cGVHD cases were reclassified
- Incidence of cGVHD was nearly double that of L-aGVHD
- Bronchiolitis obliterans (BO) defined by lung biopsy and pulmonary function testing (PFT) criteria is difficult to diagnose in young children because of their inability to adequately complete PFT and inconsistent lung biopsies, hindering the diagnosis of pulmonary cGVHD
- Of 12 reported patients with pulmonary cGVHD, 8 met formal 2014 NIH-CC criteria for pulmonary cGVHD (pathological BO or BOS)

3. According to the prospective, multi-institution study by Cuvelier and colleagues, which of the following statements about the clinical features and risk factors for cGVHD and L-aGVHD in children is correct?

- The mouth, skin, eyes, and lungs were the most common organs involved with diagnostic or distinctive manifestations of cGVHD
- Several patients had distinctive cGVHD features of the gastrointestinal tract (esophageal webs or strictures) or genitalia (lichen planus, vaginal scarring/stenosis, ulceration, erosions, fissures)
- Younger recipient age and female sex were major risk factors for cGVHD and L-aGVHD
- Only half of patients with cGVHD had involvement of ≥ 1 organ system