

# Pediatric Modification of the Montreal Classification for Inflammatory Bowel Disease: The Paris Classification

Arie Levine, MD,\* Anne Griffiths, MD,<sup>†</sup> James Markowitz, MD,<sup>‡</sup> David C Wilson, MD,<sup>§</sup> Dan Turner, MD, PhD,<sup>¶</sup> Richard K Russell, MD, PhD,<sup>||</sup> John Fell, MD,<sup>\*\*</sup> Frank M Ruemmele, MD, PhD,<sup>††</sup> Thomas Walters, MD,<sup>‡</sup> Mary Sherlock, MD,<sup>‡</sup> Marla Dubinsky, MD,<sup>‡‡</sup> and Jeffrey S Hyams, MD<sup>§§</sup>

**Background:** Crohn's disease and ulcerative colitis are complex disorders with some shared and many unique predisposing genes. Accurate phenotype classification is essential in determining the utility of genotype–phenotype correlation. The Montreal Classification of IBD has several weaknesses with respect to classification of children. The dynamic features of pediatric disease phenotype (change in disease location and behavior over time, growth failure) are not sufficiently captured by the current Montreal Classification.

**Methods:** Focusing on facilitating research in pediatric inflammatory bowel disease (IBD), and creating uniform standards for defining IBD phenotypes, an international group of pediatric IBD experts met in Paris, France to develop evidence-based consensus recommendations for a pediatric modification of the Montreal criteria.

**Results:** Important modifications developed include classifying age at diagnosis as A1a (0 to <10 years), A1b (10 to <17 years), A2 (17 to 40 years), and A3 (>40 years), distinguishing disease above the distal ileum as L4a (proximal to ligament of Treitz) and L4b (ligament of Treitz to above distal ileum), allowing both stenosing and penetrating disease to be classified in the same patient (B2B3), denoting the presence of growth failure in the

patient at any time as G<sub>1</sub> versus G<sub>0</sub> (never growth failure), adding E4 to denote extent of ulcerative colitis that is proximal to the hepatic flexure, and denoting ever severe ulcerative colitis during disease course by S1.

**Conclusions:** These modifications are termed the Paris Classification. By adhering to the Montreal framework, we have not jeopardized or altered the ability to use this classification for adult onset disease or by adult gastroenterologists.

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**Key Words:** inflammatory bowel disease, Crohn's disease, ulcerative colitis, phenotypes, children

Inflammatory bowel disease (IBD) develops during childhood or adolescence in up to 25% of patients. Although the diagnoses ulcerative colitis (UC) and Crohn's disease (CD) are applied to differentiate the two major phenotypic forms of IBD, it is recognized that both, particularly CD, comprise a spectrum of chronic intestinal inflammation, with significant variation. As the genetic basis of susceptibility to IBD has been explored, first via linkage and subsequently via genome-wide association studies, it has become clear that CD and UC are complex disorders with some shared and many unique predisposing genes. In addition to providing clues concerning disease pathogenesis, knowledge of genetic polymorphisms in predisposing and modifying genes may explain observed variations in disease location, behavior, severity, and responsiveness to therapies. Accurate phenotype classification, which captures disease type and evolution, however, is essential if the utility of genotypic data in predicting clinical variability is to be assessed. The Montreal Classification of IBD<sup>1</sup> was developed with that purpose in mind. However, the Montreal Classification has several weaknesses with respect to classification of young patients.

Descriptive data from pediatric registries around the world have highlighted phenotypic characteristics unique to pediatric IBD, including a greater propensity for disease extension. The dynamic features of the pediatric disease phenotype (i.e., change in disease location and in disease behavior over time) are not sufficiently captured by the current Montreal Classification. Linear growth impairment is not considered at all and age at onset is only arbitrarily

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From the \*Wolfson Medical Center, Tel Aviv University, Israel, <sup>†</sup>Hospital for Sick Children, Toronto, Ontario, Canada, <sup>‡</sup>Cohen Children Medical Center of New York, New Hyde Park, New York, <sup>§</sup>Child Life and Health, University of Edinburgh and Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Edinburgh, Scotland, UK, <sup>¶</sup>Shaare Zedek Medical Center, Jerusalem, Israel, <sup>||</sup>Department of Paediatric Gastroenterology and Nutrition, Royal Hospital for Sick Children, Glasgow, Scotland, UK, <sup>\*\*</sup>Imperial College, London, UK, <sup>††</sup>Université Paris-Descartes, INSERM U989, Assistance Publique-Hopitaux de Paris, Hôpital Necker Enfants Malades, Paris, France, <sup>‡‡</sup>Department of Pediatrics, Maxime Dunitz Children Hospital, Pediatric IBD Center, Cedars-Sinai Medical Center, Los Angeles, California, <sup>§§</sup>Connecticut Children Medical Center, Hartford, Connecticut.

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Reprints: Jeffrey S Hyams, MD, Connecticut Children Medical Center, 282 Washington St., Hartford, CT 06106. (e-mail: jhyams@ccmckids.org)

All authors contributed equally to this article.

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categorized. Moreover, the Montreal Classification has been recently shown to have only moderate interrater reliability when utilized in pediatric IBD, suggesting that the diagnostic criteria applied to young patients may need to be modified (M. Sherlock, unpubl. data).

With a specific focus on facilitating research in pediatric IBD, and creating uniform standards for defining IBD phenotypes, an international group of pediatric IBD experts met to develop evidence-based consensus recommendations for a pediatric modification of the Montreal criteria. This work was largely completed in conjunction with the 2nd International Symposium on Pediatric Inflammatory Bowel Disease held in Paris, France in September 2009. This new classification of pediatric IBD, the Paris Classification, reflects currently available evidence and clinical practice of pediatric IBD.

### MATERIALS AND METHODS

In May 2009 an international group of experts in pediatric IBD examined the literature to determine whether and how specific domains of the Montreal Classification required modification to reflect the phenotypic spectrum of pediatric IBD. In particular, the objectives were:

1. to classify age of onset into appropriate categories based on analysis of variation in phenotypic spectrum of IBD according to age of onset;
2. to provide definitions of CD versus UC versus IBD-U that reflect reported observations in younger patients;
3. to optimally classify location of pediatric CD and UC;
4. to derive pediatric definitions of disease behavior;
5. to incorporate disease duration into descriptions of disease behavior and location;
6. to define and standardize disease classification;
7. to classify growth as normal or impaired.

### Approach to Evidence Review

The group met three times during the course of 2009. In an initial meeting of the consensus panel (May 2009), each of the above objectives was assigned to a subgroup of two to three members to draft an initial document based on a literature review. A uniform search strategy was used to identify relevant pediatric IBD studies including: clinical guidelines, systematic reviews and meta-analyses, clinical trials, other controlled trials, cohort studies, case-control studies, diagnostic studies, case series, and expert opinion (including narrative review). Particular emphasis was placed on identifying and including studies that analyzed phenotypes by age (annualized data) within the pediatric age group. Whenever data were available we evaluated the effect of age, macroscopic and microscopic involvement on disease phenotype, and behavior and serological response. These findings were then discussed at meetings held in

Paris, France and Hollywood, Florida. Areas of consensus and need for reconsideration emerged and subsequently a consensus document was prepared after final approval by all participants.

## RESULTS

### Type of IBD (UC Versus CD Versus IBD-U)

Differentiation of UC from CD, and the use of the label IBD-U, were the subject of a previous Working Group on the classification of pediatric IBD.<sup>2</sup> The focus of that systematic review was to rigorously review evidence concerning controversial issues in the labeling of patients (usually with predominantly colonic disease) as CD or UC or IBD-U. These included upper tract macroscopic and microscopic inflammation or short segment ileal inflammation in patients with continuous pancolitis. This committee re-reviewed the evidence and supported the consensus recommendations as summarized in that report.

Macroscopic rectal sparing may be seen in about 10%–30% of children and adults with UC, most of whom have “relative rectal sparing” (mild patchy disease) rather than “absolute macroscopic rectal sparing.”<sup>3–6</sup> Microscopic evidence of chronicity in the presence of acute inflammation is typical of UC; however, histological signs of chronicity can be absent at presentation. This may reflect a shorter duration of symptoms before biopsy in children.<sup>7</sup> The presence of even one well-formed noncaseating granuloma remote from ruptured crypts anywhere in the gastrointestinal tract should prompt the diagnosis of CD. The presence of focal active colitis is not consistent with untreated UC. In a study of 29 children with this finding at diagnosis only one (3%) was subsequently diagnosed as UC.<sup>8</sup> Microscopically normal appearing skip lesions should preclude the diagnosis of UC. A patch of inflammation surrounding the appendix (called “cecal patch”) may be commonly seen in UC with only left-sided inflammation.<sup>9</sup>

Microscopic rectal sparing (i.e., absolute histological rectal sparing) is uncommon in UC.<sup>10,11</sup> In children, only 2/73 (3%) and 2/30 (7%) newly diagnosed patients had complete histological rectal sparing.<sup>3,12</sup> Some of these patients prove to have CD years after the initial diagnosis.<sup>4</sup> Microscopically normal-appearing rectum should, therefore, lead to the diagnosis of IBD-U or CD depending on other findings. A summary of factors that preclude a diagnosis of UC is shown in Table 1.

Mild, nonspecific mucosal changes in the upper gastrointestinal tract are very common in both CD and UC patients<sup>2</sup> but multiple ulcerations in the esophagus, stomach, or duodenum are rarely seen in UC (0%–8%).<sup>2,13,14</sup> Extensive macroscopic inflammation of the upper gastrointestinal tract, in particular serpentine ulcers and cobblestoning, should prompt the diagnosis of CD. Histological upper

**TABLE 1.** Consensus on Features that Preclude Diagnosis of Ulcerative Colitis

Presence of perianal disease (as defined)
Microscopically normal appearing skip lesions
Microscopically normal appearing rectum (i.e., absolute histologic rectal sparing)
Stenosis, cobblestoning, and linear ulcerations in the ileum (even in the presence of pancolitis)
Any macroscopic ileitis in the presence of normally looking cecum
The presence of even one well-formed granuloma remote from ruptured crypts
Extensive macroscopic inflammation of the UGI tract (e.g., serpentine ulcers and cobblestoning)

gastrointestinal changes alone should not exclude the diagnosis of UC<sup>15–18</sup> except in the presence of granuloma.

Backwash ileitis may occur in up to 20% of UC patients with pancolitis.<sup>10,19–21</sup> Stenosis, cobblestoning, and linear ulcerations in the ileum, or inflamed ileum with a normal cecum, are not compatible with backwash ileitis and should prompt the diagnosis of CD. Submucosal inflammation, or crypt architectural abnormality or atrophy, are not typical of backwash ileitis and should lead to the diagnosis of IBD-U or CD.<sup>20–22</sup> A few small ulcerations in the small bowel found on capsule endoscopy do not preclude the diagnosis of UC, since these may be found in a significant proportion of healthy individuals, and also since some degree of nonspecific small bowel inflammation can be found in UC.

### Age of Onset

The Montreal Classification defined three age categories (A1 ≤16 years, A2 17–40 years, A3 >40 years). The pediatric cutoff of ≤16 was based on the need for a pediatric age group but was not evidence-based.

In CD, location or extent of disease differs according to age of disease onset. Very early age of onset is characterized predominantly by isolated colitis, with ileal disease occurring more often in children whose disease is diagnosed after the age of 9–10 years.<sup>23–26</sup> This colitis predominant phenotype is most prominent, but not limited to, children without *NOD2* mutations.<sup>25,26</sup> Thus, evidence suggests an appropriate cutpoint for age of 9–10 years based on observed variation in the phenotypic spectrum.<sup>25,26</sup>

Variation in serologic responses by age is similarly supportive of a classification that distinguishes IBD onset as before or after 9 years of age.<sup>27</sup> The rate of detectable anti-*Saccharomyces cerevisiae* antibody (ASCA) titers increases significantly from the age of 8 years and it appears to reach a steady state between 10–15 years. The

percentage of children with detectable anti-CBir1 is highest during the first few years of life, and declines with increasing age at disease diagnosis.<sup>27</sup> In contrast, the presence of perianal disease and disease behavior do not differ with respect to age of onset in most studies.<sup>28–32</sup>

In UC, age at diagnosis appears to affect the risk for surgery or biological therapy within the first few years after diagnosis, based primarily on an analysis performed on patients enrolled in the Pediatric Inflammatory Bowel Disease Collaborative Research Group Registry (J. Markowitz et al, personal communication). The rate for colectomy or biological therapy in UC was 7% in children younger than 8 years at disease onset, rose to 14% for those 8–10 years of age, and increased to 21%–30% thereafter. The most appropriate age cutoff based on maximal significance was <9 years ( $P = 0.0043$ ).

There is some evidence to subdivide a separate group of children diagnosed with IBD at a very early age (0–2 years, infantile IBD). This subgroup was more likely to have a family history in first-degree relatives (44% in the 0–2 years age group versus 19% in the 3–16 years group;  $P = 0.0002$ ), lending support to the hypothesis of a higher genetic load.<sup>34</sup> Children in this age group have more severe disease course and a high rate of resistance to immunosuppressive treatment.<sup>33,34</sup> The suspicion of a monogenetic cause of these early onset forms was recently confirmed by the discovery of mutations in the genes coding for one of the two IL10 receptors causing impaired IL10 signaling.<sup>35</sup> These findings indicate a new view of IBD as a continuum in physiopathogenic mechanisms from severe inactivating mutations causing early onset disease to gene polymorphisms that may decrease the efficacy of important immunoregulatory pathway in individuals starting disease later in life. Further research is required to determine if there is a need for a separate descriptor for infantile IBD.

Taken together, it appears that there are enough data to support an age cutoff of <10 years for classifying both CD and UC (Table 2). It is therefore our recommendation to subdivide the Montreal A1 classification into A1a, which represents 0–9 years of age, and A1b, which represents ages 10 through 16 years.

### Disease Location

#### Crohn's Disease

The frequent performance of upper gastrointestinal endoscopy in newly diagnosed pediatric patients and the increasing use of video capsule endoscopy, magnetic resonance (MR) enterography, and computed tomography (CT) enterography have increased awareness that disease proximal to the ileum is common. Classification of disease must include location but definition of location must be able to reflect all combinations of involvement. Moreover,

**TABLE 2.** Montreal and Paris Classifications for Crohn's Disease

	Montreal	Paris
Age at Diagnosis	A1: below 17 y A2: 17-40 y A3: Above 40 y	A1a: 0-<10y A1b: 10-<17 y A2: 17-40 y A3: >40 y
Location	L1: terminal ileal ± limited cecal disease L2: colonic L3: ileocolonic L4: Isolated upper disease*	L1: distal 1/3 ileum ± limited cecal disease L2: colonic L3: ileocolonic L4a: upper disease proximal to Ligament of Treitz* L4b: upper disease distal to ligament of Treitz and proximal to distal 1/3 ileum*
Behavior	B1: non-stricturing non-penetrating B2: stricturing B3: penetrating p: perianal disease modifier	B1: nonstricturing nonpenetrating B2: stricturing B3: penetrating B2B3: both penetrating and stricturing disease, either at the same or different times p: perianal disease modifier
Growth	n/a	G <sub>0</sub> : No evidence of growth delay G <sub>1</sub> : Growth delay

\*In both the Montreal and Paris Classification systems L4 and L4a/L4b may coexist with L1, L2, L3, respectively.

B1 - Nonstricturing, nonpenetrating disease: uncomplicated inflammatory disease without evidence of stricturing or penetrating disease.

B2 - Stricturing disease: the occurrence of constant luminal narrowing demonstrated by radiologic, endoscopic, or surgical examination combined with prestenotic dilation and/or obstructive signs or symptoms but without evidence of penetrating disease.

B3 - Penetrating disease: the occurrence of bowel perforation, intraabdominal fistulas, inflammatory masses and/or abscesses at any time in the course of the disease, and not secondary postoperative intra-abdominal complication (excludes isolated perianal or rectovaginal fistulae).

B2B3 - Stricturing and penetrating disease: the presence of both B2 and B3 phenotypes in the same patient, either at the same moment in time, or separately over a period of time [correction made after initial online publication].

consensus over what constitutes involvement (macroscopic, microscopic) must be achieved to allow uniformity between outcome studies. The Montreal Classification allows L4 (upper tract) to coexist with L1 (ileal), L2 (colonic), or L3 (ileocolonic) (1). The Montreal Classification as it stands still has a degree of ambiguity, pertinent to both pediatric and adult phenotyping. The L4 category does not

distinguish between small intestinal disease, and esophageal or gastric disease and recent data presented by the NIDDK genetics consortium suggested the need for distinguishing the disparate disease locations currently included in L4.<sup>36</sup> Clarity in disease distribution is critical, since significant small bowel involvement may be associated with poorer outcomes, including growth failure, weight loss, and stricturing disease,<sup>37</sup> while evidence of long-term outcomes from gastroduodenal involvement are lacking. Several pediatric studies have used histologic involvement of the upper gastrointestinal (UGI) tract in their criteria for L4, without supporting evidence on its effect on phenotype and outcome.<sup>26,38,39</sup> Finally, the definition of ileal disease with cecal involvement has not been clearly defined, leading to conflicting definitions of location as L1 (as in the Vienna classification) or L3.

UGI involvement has been estimated to range from 30%–80% in children and adults with CD.<sup>40–46</sup> Recent data from the Pediatric IBD Collaborative Research Group Registry, a prospective study of newly diagnosed children ≤16 years of age, categorized UGI involvement as esophageal or gastroduodenal, and subgrouped by gross endoscopic involvement, histologic involvement, or both, based on local practice patterns.<sup>47</sup> Approximately 80% (745/932) of children with CD had UGI endoscopy, of whom five had UGI involvement only. Esophageal involvement was seen in 203 patients (27%) (macroscopic in 18%), and gastroduodenal involvement in 413 (56%) (macroscopic in 42%). Isolated oral, isolated perianal, and isolated oral and perianal disease may occur at diagnosis, but are rare.<sup>29</sup>

In CD there are insufficient data at present to suggest that isolated microscopic involvement, in the absence of macroscopic involvement, affects disease phenotype, or disease behavior over time. The only reproducible histologic finding that has been evaluated is the presence or absence of granuloma, and the data at present are conflicting. A confounding factor present in studies is that granuloma detection rises significantly with surgical specimens leading to selection bias. The best methodology for estimating risk would therefore be granulomas at presentation and subsequent disease behavior. Two studies<sup>48,49</sup> did not identify a change in disease behavior based only on the presence of granulomas at diagnosis.

We recommend that the definition of location of CD be based on macroscopic appearance of mucosal ulceration anywhere along the GI tract (with the exception of the mouth) or bowel wall thickening on radiography. The presence of mucosal erythema, and/or granularity, is not sufficient to be considered evidence of involvement. In keeping with the Porto guidelines, we recommend evaluating the whole GI tract by upper endoscopy, ileocolonoscopy, and small bowel imaging.<sup>50</sup>

In order to facilitate clear phenotyping, with maximal ability to study associations with different disease locations,



**TABLE 3.** Montreal and Paris Classifications for Ulcerative Colitis

	Montreal	Paris
Extent	E1: ulcerative proctitis	E1: ulcerative proctitis
	E2: left-sided UC (distal to splenic flexure)	E2: Left-sided UC (distal to splenic flexure)
	E3: extensive (proximal to splenic flexure)	E3: Extensive (hepatic flexure distally)
		E4: Pancolitis (proximal to hepatic flexure)
Severity	S0: clinical remission	S0: never severe*
	S1: mild UC	S1: ever severe*
	S2: moderate UC	
	S3: severe UC	

\*Severe defined by Pediatric Ulcerative Colitis Activity Index (PUCAI)  $\geq 65$ .<sup>58</sup>

we recommend the following modifications to the Montreal Classification noted in Table 2: L1 = distal 1/3 of the small intestine with limited or no cecal disease. Specifically, if there is evidence of limited cecal disease, and no evidence of other colonic involvement in patients with ileal involvement, then this should be labeled L1. L2 remains disease confined to the colon. Colonic disease beyond the cecum with ileal involvement remains L3 as per Montreal. Similarly, if the colon is normal except for the presence of a fistula extending from inflamed small bowel, the patient should be defined as L1 only. We elected to modify L4 and to separate gastroesophageal or duodenal disease from jejunal/proximal ileal disease in the following fashion: L4a is defined by disease proximal to the ligament of Trietz, and L4b is disease distal to the ligament of Trietz but proximal to the distal third of the small intestine. An individual could be both L4a and L4b. Disease location should be defined by macroscopic findings and not by histology in otherwise healthy appearing mucosa.

### Ulcerative Colitis

In the Montreal Classification, disease extent for UC was divided into three categories. The first category (E1) describes patients with proctitis, (E2) patients with left-sided disease distal to the splenic flexure, and the last category (E3) describes patients with extensive disease proximal to the splenic flexure. Disease extent is defined in the classification using macroscopic appearance rather than evidence from histology/radiology. Pediatric-onset UC is characterized by extensive colitis or pancolitis in the majority of cases.<sup>24,29,51,52</sup> A greater risk of colectomy with more extensive disease has been noted in previous adult studies.<sup>52,53</sup> In order to delineate the importance of extensive disease in future studies, we recommend adding a new cat-

egory (E4) for pancolitis (disease extending from rectum to proximal to the hepatic flexure). Our recommendations for classification of UC disease location are shown in Table 3.

### Disease Behavior

#### Crohn's Disease

The Montreal Classification defines three behaviors for CD: nonstricturing nonpenetrating disease (B1), stricturing disease (B2), penetrating disease (B3). Perianal and rectovaginal fistula(s) without additional evidence of fistulizing disease are not defined as B3. To date, patients with evidence of both stricturing and penetrating disease are classified as B3.

Observations drawn from cohort and large cross-sectional studies suggest that children and adults with CD do not have significantly different CD behavior, either at the time of diagnosis or over time.<sup>28,29,31</sup> Approximately 20% of adults present with penetrating or stricturing CD at diagnosis<sup>54</sup> but the rate increases over time, such that the 20-year actuarial rate for persistent inflammatory phenotype remains only 12%. In a prospectively enrolled North American pediatric cohort, only 12% of 796 children had B2 or B3 disease at diagnosis, but another 20% developed these behaviors during a median follow-up of 32 months.<sup>55</sup> Similar CD behavior was observed in two additional pediatric studies. In one describing Scottish children with CD, 91% were B1 at diagnosis while 24% developed B2 or B3 within 4 years of diagnosis.<sup>29</sup> In the French EPIMAD study, with a longer follow-up (median follow-up of 7 years), complicated CD behavior (B2 and B3) doubled during the follow-up period from 29% at diagnosis to 59%, reaching a relative plateau after 9–10 years.<sup>32</sup>

The Montreal Classification defines subjects with evidence of both stricturing and penetrating disease as B3, largely based on the article by Oberhuber et al.<sup>56</sup> Using surgical resection specimens, that study localized 26/27 fistulae within or at the proximal end of a stricture, and identified only one fistula not associated with a stricture. By contrast, a pediatric study<sup>57</sup> reported 28 specimens with fistulae, of which only 16 had associated stenotic disease. In these 16 specimens, seven fistulae were localized proximal to a stricture, five distal to a stricture, and four specimens contained fistulae both proximal and distal to a stricture. It therefore appears that penetrating and stricturing CD can be independent of one another and at times coexist. Data from the aforementioned North American pediatric database reveals that 34 of the 780 (4.4%) children presenting with B1 behavior at the time of diagnosis had both stricturing and penetrating disease behavior at a mean follow-up of 32 months (unpubl. data in Ref. 55).

Based on these data, we propose a new classification B2B3 that should be used to identify individuals with both

**TABLE 4.** Suggested Classification Paradigm for Documenting Linear Growth Impairment in Pediatric Onset IBD

	Definition
$G_0$	Normal growth at diagnosis and subsequently (i.e., not meeting any of the definitions of growth impairment as defined below)
$G_1$	Impaired linear growth as defined by at least one of the following criteria <ol style="list-style-type: none"> <li>1) Height z-score at diagnosis or subsequently significantly less than expected height z-score               <ol style="list-style-type: none"> <li>A) Difference between observed height z-score and predicted height z-score using the 'Mid-parental Heights' formula is <math>&gt;2.0</math> OR B) Difference between observed height z-score and the 'pre-illness' height z-score is <math>&gt;1.0</math></li> </ol> </li> <li>2) Current height z-score significantly less than height z-score at diagnosis Reduction in height z-score since diagnosis is <math>\geq 0.75</math></li> </ol>

behaviors (developing either concomitantly or serially) (Table 2). The B2B3 classification will allow such individuals to be more easily distinguished from those who develop fistulizing disease without associated stricturing of the bowel. Despite lack of our consensus regarding the importance of violaceous tags, we recommend to continue using the descriptor p for perianal disease only if fistula, anal canal ulcers, or abscess are present. At this point we do not recommend subcategorization by other extraintestinal manifestations such as frank arthritis, uveitis, pyoderma gangrenosum, or metastatic CD.

### Ulcerative Colitis

To the best of our knowledge, no studies have specifically explored a disease behavior classification for pediatric patients with UC. The Montreal Classification summarized UC activity in terms of severity, and graded the severity of acute relapse (S0-S3). The Pediatric Ulcerative Colitis Activity Index (PUCAI) has allowed objective clinical definition of disease severity in pediatric cases.<sup>58</sup> The outcome of severe disease has been reported in relation to progress to second-line therapy and colectomy rate following initial intravenous corticosteroid therapy in a retrospective cohort study of 99 children<sup>59</sup> and a prospective multicenter study of 128 children.<sup>60</sup> Specifically, those with high PUCAI scores at the beginning of the index admission for intravenous corticosteroids had significantly higher colectomy rates during the following year in both cohorts.

In Table 3 we adopted a disease behavior classification as S0 or S1, with the latter denoting the presence of

severe disease at any time in the patient history (as previously defined by a PUCAI score of  $\geq 65$ ).

### Disease Duration and Effects on Location, Behavior, and Disease Severity

Since disease extent is rather stable in adult onset CD, the issue of timing of assessment is relevant mainly for disease behavior.<sup>61</sup> In contrast, a significant change in both disease location and behavior were noted over time in a large cohort of pediatric patients with CD.<sup>29</sup> Unlike in adults, CD location was dynamic in childhood-onset disease; within 2 years of diagnosis, childhood-onset CD progressed to involve additional sites in 39% of patients who did not already have the maximal disease extent (L3 and L4). CD behavior changed with a decrease in inflammatory disease (B1) from 91.2% to 82.7% by 2 years and 75.8% by 4 years. In another pediatric study of 404 patients with childhood-onset CD and a median follow-up of 7 years,<sup>32</sup> it was shown that complicated disease behavior (B2 and B3) doubled during the follow-up period from 29% at diagnosis to 59%, reaching a relative plateau after  $\approx 9$ –10 years.

In pediatric UC, extensive disease location changes over time.<sup>29</sup> In one multicenter study involving 38 cases of childhood-onset ulcerative proctitis, proximal extension occurred in 29% during follow-up ranging from 6 months to 11 years; over 50% of subjects followed for more than 5 years had proximal extension.<sup>62</sup> In another cohort of 113 children with new onset UC followed for a median of 6 years, the rate of extensive disease increased from 37% to 60% at last follow-up.<sup>51</sup>

We therefore recommend performing regular updates of IBD location and behavior throughout the course of IBD in children and young people. Reports of disease behavior should be described in relation to disease duration.

### Growth

As growth abnormalities are an important element of disease phenotype in pediatric IBD,<sup>63</sup> the committee felt that growth must be included in the phenotypic classification. Growth abnormalities at presentation are noted in up to 30% of children with CD; however, studies have not used uniform definitions of abnormal growth.<sup>64</sup>

The adequacy of growth over a specified time frame is best characterized by linear growth velocity, standardized for gender, age, and pubertal development, using time-points as close to 12 months apart as possible and using standardized scores (z-scores) for height. Different metrics are required to diagnose growth retardation at diagnosis or during follow-up.

A height velocity z-score of approximately  $-2$  over a 12-month period equates to a reduction in height z-score of approximately 0.3 to 0.4. By extrapolation, a subject who

experiences persistent linear growth delay with height velocity z-scores below the 3rd percentile (z-score  $-2$ ) for at least 2 years would demonstrate a reduction in height z-score in the range of about 0.75. In the absence of other guidelines, it would seem reasonable to define a reduction in height z-score (since diagnosis) of  $\geq 0.75$  as evidence of postdiagnosis linear growth impairment.

Accurate documentation of pubertal development along with the evaluation of radiological bone age are crucial for growth evaluation. Females enter the adolescent growth spurt relatively early in puberty, while in males it occurs in late puberty (Tanner 4).<sup>65</sup> The growing phase could be considered final once they have entered Tanner stage 5 and they have demonstrated less than 0.5 cm linear growth in 12 months.

Table 4 shows our recommended classification for documenting normalcy or impairment of linear growth in children with IBD.  $G_0$  defines normal growth at diagnosis and subsequently, whereas  $G_1$  denotes a height z-score at diagnosis or subsequently that is significantly less than the expected height z-score

## CONCLUSIONS

We have modified and modernized the Montreal Classification to facilitate standardization of definitions of disease phenotype based on the best pediatric evidence and other changes in practice that have happened since the guidelines were developed in 2005. We recognize that the Vienna and Montreal Classifications acted as benchmarks that have been adapted over time as studies have highlighted the benefits and deficiencies with them. We envisage the same process will happen with adoption of the Paris Classification. By adhering to the existing Montreal framework, we have not jeopardized or altered the ability to use this classification for adult onset disease or by adult gastroenterologists. It is our hope that this classification could be used by both pediatric and adult gastroenterologists to facilitate simplicity and uniformity for future studies.

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