Inflammatory Bowel Disease in Children & Adolescents

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“The 100-calorie” IBD Lecture

“IBD-Lite”

…and now for an IBD Commercial

Epidemiology of IBD in Children & Adolescents

Crohn’s Disease rules.
Crohn’s Disease - 60%
Ulcerative Colitis - 30%
Indeterminate Colitis - 10%

Typically presents 2nd decade of life.
Inflammatory Bowel Disease
Immunologic Dysfunction

- Genetic Predisposition
- Environmental Triggers
- Immune Abnormalities
- Tissue Injury

Crohn’s Disease: Pathology

Categorization based on disease behavior
- **Inflammatory**
  - transmural, typically patchy and characterized by granulomas
- **Stricturing**
- **Penetrating** (fistulizing)

Disease distribution
- Occurs anywhere mouth to anus
- Ileocolonic disease occurs in 50%
- Perianal disease occurs in 50%

Endoscopy in CD

Individual Ulcers
Ulcerative Colitis

Inflammation: involves the rectum and extends proximally in an uninterrupted pattern to involve all or part of the large intestine

- Proctitis - 15%
- Left sided colitis - 25%
- Total colonic disease - 60%

Michaels, Caulfield, Wyllie et al. 1990

Endoscopy in UC

Diffuse Mucosal Inflammation

Presenting Features in Childhood

<table>
<thead>
<tr>
<th></th>
<th>CD</th>
<th>UC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal Pain</td>
<td>51%</td>
<td>5%</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>62%</td>
<td>53%</td>
</tr>
<tr>
<td>Weight Loss</td>
<td>93%</td>
<td>10%</td>
</tr>
<tr>
<td>Fever</td>
<td>22%</td>
<td>-</td>
</tr>
<tr>
<td>Rectal Bleeding</td>
<td>23%</td>
<td>20%</td>
</tr>
<tr>
<td>Arthritis</td>
<td>9%</td>
<td>4%</td>
</tr>
<tr>
<td>Growth Failure</td>
<td>40%</td>
<td>2%</td>
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</tbody>
</table>

Calenda and Grand 1995
Extraintestinal Manifestations of IBD in Children

<table>
<thead>
<tr>
<th></th>
<th>CD</th>
<th>UC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Joints</td>
<td>11%</td>
<td>14%</td>
</tr>
<tr>
<td>Eyes</td>
<td>0.2%</td>
<td>1-2%</td>
</tr>
<tr>
<td>Skin</td>
<td>7-13%</td>
<td>1-2%</td>
</tr>
<tr>
<td>Liver</td>
<td>?</td>
<td>1-2%</td>
</tr>
<tr>
<td>Fistulas</td>
<td>15%</td>
<td>0</td>
</tr>
<tr>
<td>Renal Calculi</td>
<td>5%</td>
<td>0-5%</td>
</tr>
</tbody>
</table>

Calenda & Grand 1995

Extraintestinal Manifestations

Skin
- Erythema nodosum (4%)
  - lower extremity: typically over shins
- Pyoderma gangrenosum (1%)
  - lower extremity typically over ankles
- Other skin conditions
  - Polyarteritis nodosa
  - Vasculitis
  - Epidermolysis bullosa

Erythema Nodosum (4%)
Pyoderma Gangrenosum (1%)
Extraintestinal Manifestations

Ocular

- Ocular complications (< 10%)
  - Episcleritis
  - Iritis
  - Uveitis
  - Orbital pseudotumor
- Subcapsular cataracts - steroids

Gastrointestinal Complications

- **Hemorrhage** - massive 1-3% pts with IBD
- **Intestinal obstruction** - usually partial small bowel in CD; colonic strictures in UC
- **Perforation** - localized in CD; toxic megacolon in UC
- **Abscess** - with fever and abdominal pain; more common in CD
- **Toxic megacolon** - acute dilation of the colon; more common in UC. Treated with antibiotics, decompression and/or surgery. Avoid agents that slow motility in severe IBD.

Inflammatory Bowel Disease Diagnosis

- **History + PE + laboratory evaluation + appropriate radiologic, endoscopic and histologic findings = DIAGNOSIS**
- **Clinical History**
  - Abdominal pain, diarrhea, hematochezia
  - Fever, fatigue, weight loss
  - Extra-intestinal sx: joint complaints, rash, etc.
  - Family history of IBD
Inflammatory Bowel Disease

Physical Examination

General appearance: pallor, fatigue, nutritional assessment (review growth chart data)

HEENT: conjunctivae, oral ulcers
Abdomen: BS, tenderness, mass
Rectal: perianal disease; heme+/-
Extremities: clubbing, joint deformity/ROM
Skin: rashes

Growth Chart in IBD

Crossing Channels

Inflammatory Bowel Disease

- Laboratory Evaluation
  - Anemia - 70%
  - Elevated sed rate - 80%
  - Hypoalbuminemia - 60%
  - Thrombocytosis - 60%
  - Occult blood in stool - 35%
Inflammatory Bowel Disease
Laboratory Evaluation

- Antibodies
  - pANCA (perinuclear antineutrophil cytoplasmic ab)
    - 80% of pts with UC
    - 45% of pts with CD
  - ASCA (anti-Saccharomyces cerevisiae ab)
    - 60-70% pts with CD
    - 8-14% pts with UC
  - OmpC (ab to E coli outer membrane porin C)
    - Associated with complicated small bowel disease
  - Anti-CBir1 (CD), NSNA DNAse sensitivity (UC)

Genetics

- More than 50 distinct susceptibility loci for IBD identified
- IBD1 gene on chromosome 16 encodes the protein NOD2 (CARD15); mutations confer susceptibility to ileal CD
- Loci on chromosomes 5q31-33 (IBD5), 20q13, and 21q22 have been associated with early onset CD (ie, before age 15 years)
- Loci on chromosomes 7q22 and 22q13 have been associated with UC

Radiologic Evaluation

- UGI & SBFT
  - most common abnormality: TI nodularity
  - other findings: narrowing/separation of bowel loops
- CAT scan
  - useful in identifying bowel wall thickening or abscess
- MRI enterography
  - increasingly used for imaging small bowel and extraintestinal findings (eg, perianal fistula)
  - no radiation
UGI & SBFT in IBD
Terminal Ileal Inflammation - Arrow

CT enterography

Barium Enema in UC
Loss of Haustral Folds
Endoscopy

- permits direct observation + histologic sampling
- Visually and histologically normal rectum eliminates UC
- Segmental colonic involvement, ileal disease or granulomas on biopsy establish a diagnosis of CD
- macroscopically normal bowel - may demonstrate severe histologic changes

Video Capsule Endoscopy

- Indicated in the evaluation of hemoccult positive stool/evaluation of possible small bowel Crohn’s disease
- Risk: partial or complete small bowel obstruction

Therapy

- Sulfasalazine
- 5-ASA
- Antibiotics
- Corticosteroids
- Antimetabolites
  - Azathioprine/6-MP
  - Methotrexate
- Cyclosporine
- Biologic therapy (Infliximab, Adalimumab)
Corticosteroids

- Recommended for pts that do not respond to aminosalicylates or moderate to severe disease
- Response rate
  - UC 45-90%
  - CD 53-78%
- Not effective in maintaining remission
- Side effects are dose dependent
  - Alternate day less side effects than daily

Corticosteroids
Side Effects

<table>
<thead>
<tr>
<th>Cosmetic</th>
<th>Chronic Administration</th>
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<tbody>
<tr>
<td>Acne</td>
<td>Growth suppression</td>
</tr>
<tr>
<td>Facial puffiness</td>
<td>Cataracts</td>
</tr>
<tr>
<td>Hirsutism</td>
<td>Glaucoma</td>
</tr>
<tr>
<td>Striae</td>
<td>Hypertension</td>
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<tr>
<td></td>
<td>Osteoporosis</td>
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<td></td>
<td>Myopathy</td>
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Immune Modulators
Azathioprine & 6-MP

- Immune modulators: used in conjunction with other drugs to reduce or eliminate the use of steroids
- Response rate
  - UC - ¼ pts (29-78%)
  - CD - 2/3 pts (36-91%)
- Onset of action is 3-4 months
- Effective as maintenance medications
- Complications include: pancreatitis, marrow suppression and liver toxicity

Stein and Hanauer: Gastro Clin N Am 1999
Surgery for IBD

- Surgery is curative in UC: ileoanal pouch anastomosis is curative with a high patient satisfaction rate - 157 children from CCF (> 1000 adult pts)
- Surgery in CD: not curative

Question 1.

All the following are true about Crohn’s disease except:

A. Inflammation may involve any part of the GI tract
B. Growth failure is common
C. Surgery is curative
D. It is the most common form of IBD in children.
Question 2.

Maintenance therapy for IBD includes all of the following drugs except:

A. Mesalamine
B. Corticosteroids
C. Azathioprine (Imuran)
D. Methotrexate

Question 3.

Common complications of prednisone include which of the following:

A. Growth failure
B. Abnormal liver functions
C. Osteopenia
D. Cardiomyopathy
E. Both A and C
Question 3.
A. Growth failure  
B. Abnormal liver functions  
C. Osteopenia  
D. Cardiomyopathy  
E. Both A and C

Question 4.
Which of the following are characteristic of children with ulcerative colitis?

A. Perianal disease  
B. Hematochezia  
C. Granulomas on endoscopic biopsy specimens  
D. Melena  
E. Both A and C