Sociedad Argentina de Pediatría
Jornadas Nacionales del Centenario
Gastroenterología, Hepatología y Nutrición Pediátricas

- **Caso Clínico N° 10**

Dr. Gonzalo Ortiz
Sección Gastroenterología Infantil
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Valeria

- **Edad**: 5 años
- **MC**: Deposiciones con moco y sangre, dolor abdominal, pujos, tenesmo de 1 mes de evolución.
- **Antec. personales**:

  Episodios de diarrea desde los 3 años.

  Perdida de peso en los últimos tres meses.
- **No presentaba antecedentes familiares de importancia.**

Examen Físico

- Hemodinámicamente compensada
- Palidez de piel y mucosa
- Abdomen distendido
- Dolor difuso a la palpación
- Se palpa lóbulo izquierdo de hígado a 2 cm del RC.
- Disminución de TCS
- Peso 16 kg **Pc 10** y talla 104 cm **Pc 10**

Hallazgos

- Colitis.
- Retraso de crecimiento.
- Hepatomegalia.

Exámenes complementarios

- **Laboratorio**:
  - Hto 31%
  - Hb: 11,1 g %
  - Plaq: 512000 mm^3
  - ERS: 13 mm/h
  - BT: 0.3 U/l
  - TGO: 764 U/l
  - TGP: 511 U/l
  - FAL: 2260 U/l
  - GGT: 395
  - TP: 101%
  - Proteínas totales: 8,6 g %
  - Albúmina: 3,6 g %
  - γ: 2,67 g %
  - Coprocultivo: Neg
  - PMF: Neg
  - Serologias:
  - HBs Ag (-)
  - HA V (-)
  - HIV (-)
  - HVC (-)

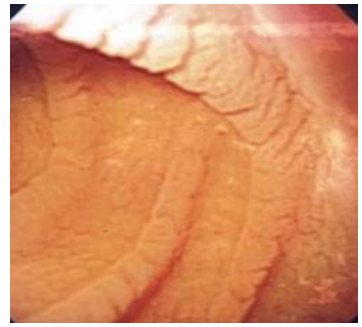
Exámenes complementarios

- **<u>Ecografía Abdominal</u>**:

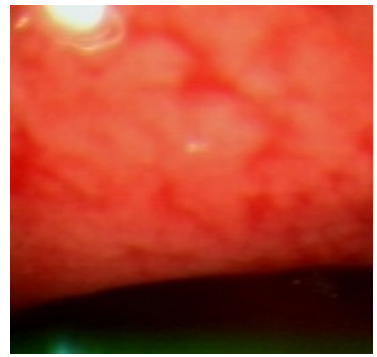
  Hígado: homogéneo, ligeramente aumentado de tamaño, vía biliar no dilatada.

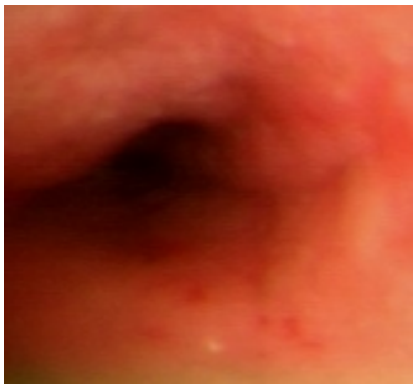
  Resto S/P.

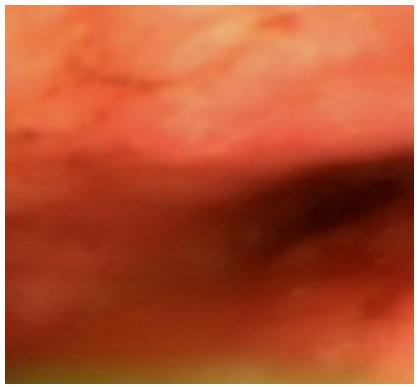
- **VEDA**: mucosa esofágica y gástrica de aspecto normal, mucosa duodenal en mosaico, pliegues con festón.



- **VCC**: se progresa hasta ciego, mucosa con eritema, edema, úlceras cubiertas con fibrina en toda su extensión. (pancolitis)







Se solicitan Anticuerpos:

- **p-Anca: +** debil
- **AGA:**
  - **IgA: 57 U/l**
  - **IgG: 52 U/l**
- **EMA: IgA +++**
- **anti-TtG: IgA >100 U/l**

- Por persistencia de hipertransaminasemia

  Se solicitan:

  - **Anticuerpos:** FAN: (-), AML (-), AntiLKM (-)

  - **Colangioresonancia**:
    - Vesícula Biliar distendida
    - Irregularidades de paredes a nivel ductal del árbol biliar, compatible con colangitis esclerosante.

  - **PBH**

Diagnósticos presuntivos

- Enfermedad Celiaca.
- EII
- Colangitis Esclerosante Primaria.

Se inicia tratamiento con:

- Dieta libre de gluten.
- Mesalazina 50 mg/kg/día.
- Acido ursodesoxicólico a 20 mg/kg/día.

Anatomía Patológica

- **Duodeno**: Histoarquitectura alterada, atrofia subtotal de vellosidades con elongación de criptas, aumento de LIES. Marsh IIIB.
- **Colon**: Severa recto colitis activa ulcerada, criptitis y abscesos crípticos aislados, compatible con colitis ulcerosa.
- **Hígado**: Hepatopatía crónica con escasa actividad inflamatoria intraacinar, fibrosis portal, y proliferación ductular. Compatible con colangitis esclerosante.

Evolución

Al mes del tratamiento, persiste con distensión abdominal, deposiciones desligadas, sumándose compromiso articular en manos y tobillos.

Meprednisona 40 mg/m^2
Azatioprina 1,5 mg/kg/día

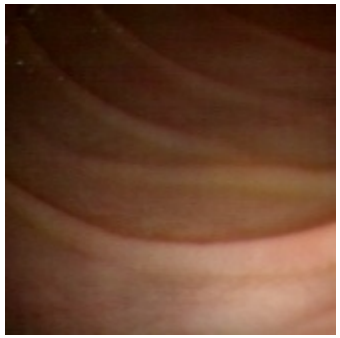
Evolución

- **Al tercer mes** del tratamiento mejoran los parámetros clínicos y del laboratorio.

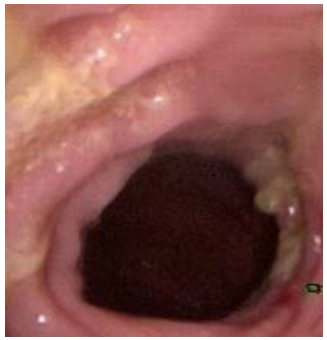
• Se inicia descenso de corticoides en forma paulatina hasta la suspensión al año.

Control Endoscópico:

- **VEDA:** Duodeno mucosa de aspecto normal.



- **VCC:** se progresa hasta ciego, calibre conservado, mucosa con eritema en forma difusa.



- **Anatomía Patológica**:

  Duodeno: mucosa con histoarquitectura conservada.

  Colon: ligero aumento de infiltrado inflamatorio, leve distorsión criptica, no abscesos.

En la actualidad, la paciente se encuentra asintomática.

- Continua con:

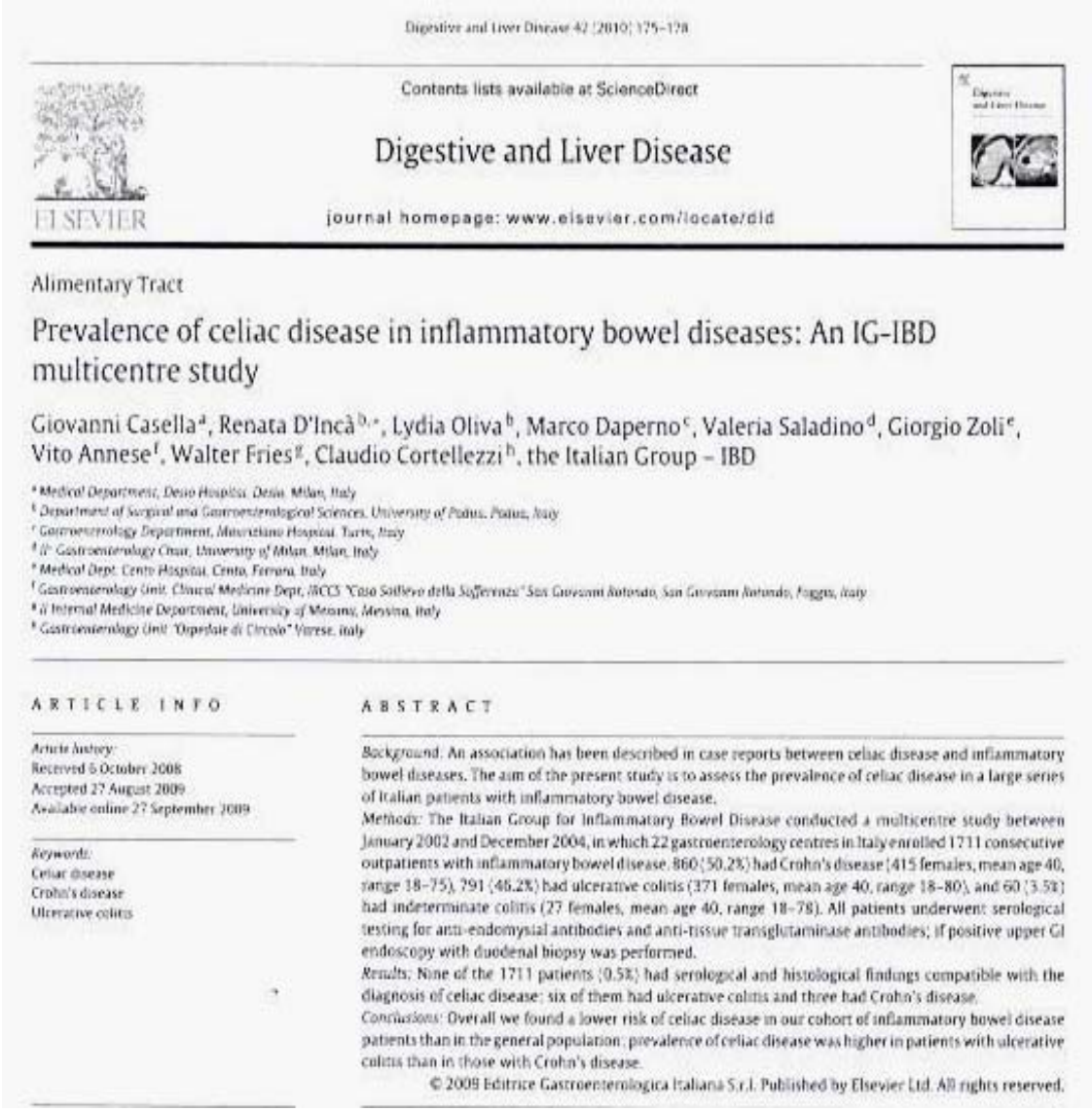
  mesalazina, acido ursodesoxicólico, azatioprina y dieta libre de gluten.

Bibliografía:

- Prevalence of celiac disease in inflammatory bowel diseases: An IG-IBD multicentre study. Claudio Cortellezzi; colab; Digestive and Liver disease; 42; 2010 175-178.
- Unique features of primary sclerosing cholangitis in children. Vergani, Current Opinion in Gastroenterology; 2010; 26:265-268.
- Hepatopanceatobiliary manifestations and complacations asociated with inflammatory bowel disease; Navaneethan U, Inflamm Bowel Dis. N°9 2010.

Muchas Gracias

Bibliografía



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Alimentary Tract

Prevalence of celiac disease in inflammatory bowel diseases: An IG-IBD multicentre study

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Celiac disease
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Ulcerative colitis

ABSTRACT

Background: An association has been described in case reports between celiac disease and inflammatory bowel diseases. The aim of the present study is to assess the prevalence of celiac disease in a large series of Italian patients with inflammatory bowel disease.

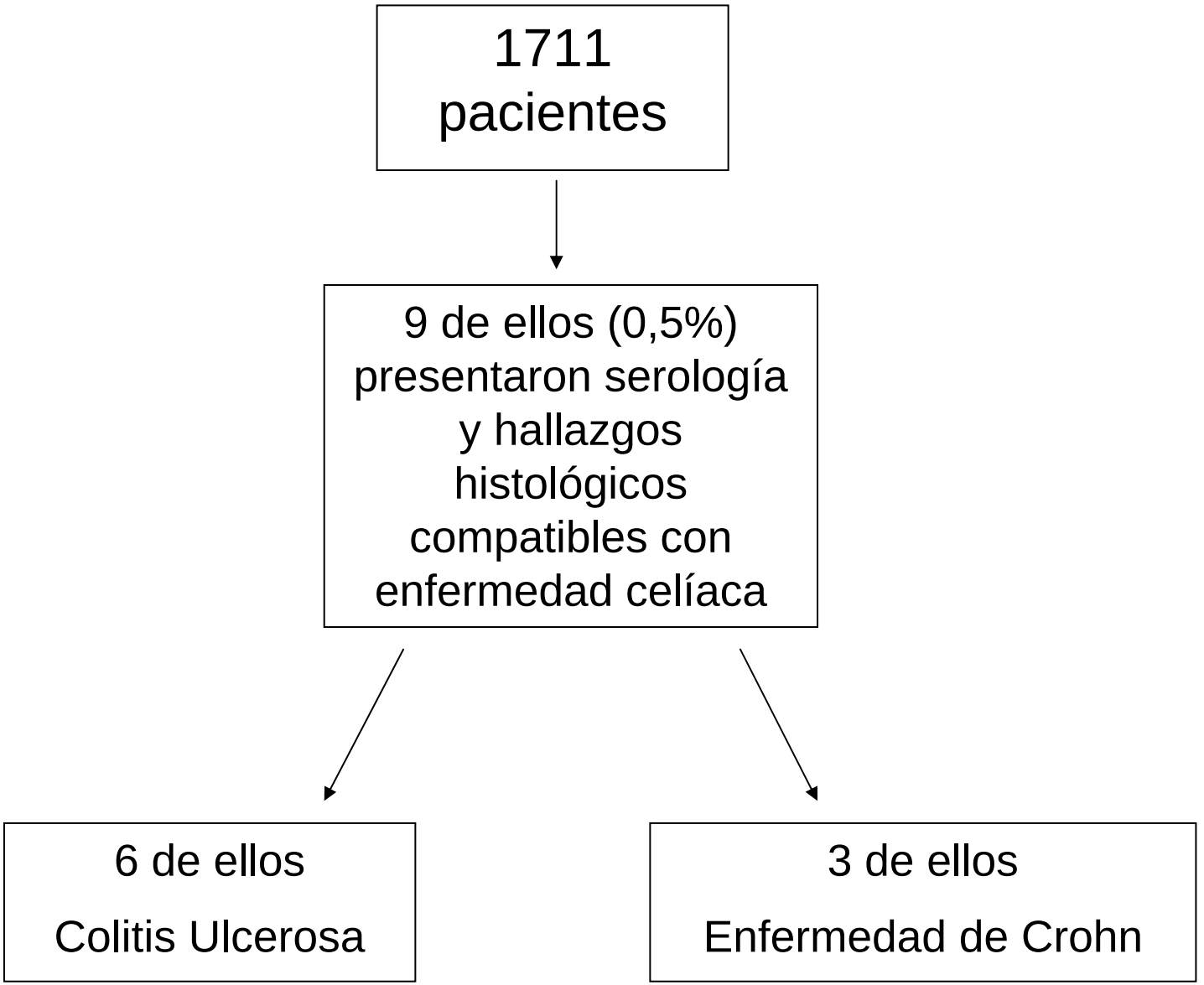
Methods: The Italian Group for Inflammatory Bowel Disease conducted a multicentre study between January 2002 and December 2004, in which 22 gastroenterology centres in Italy enrolled 1711 consecutive outpatients with inflammatory bowel disease. 860 (50.2%) had Crohn's disease (415 females, mean age 40, range 18–75), 791 (46.2%) had ulcerative colitis (371 females, mean age 40, range 18–80), and 60 (3.5%) had indeterminate colitis (27 females, mean age 40, range 18–78). All patients underwent serological testing for anti-endomysial antibodies and anti-tissue transglutaminase antibodies; if positive upper GI endoscopy with duodenal biopsy was performed.

Results: Nine of the 1711 patients (0.5%) had serological and histological findings compatible with the diagnosis of celiac disease: six of them had ulcerative colitis and three had Crohn's disease.

Conclusions: Overall we found a lower risk of celiac disease in our cohort of inflammatory bowel disease patients than in the general population: prevalence of celiac disease was higher in patients with ulcerative colitis than in those with Crohn's disease.

- Estudio Multicéntrico de Enero 2002 a Diciembre 2004

- 1711 pacientes con enfermedad inflamatoria intestinal (Crohn 860; Colitis Ulcerosa 791; Colitis indeterminada 60)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Colitis Ulcerosa
3 de ellos
Enfermedad de Crohn

Conclusión

- Se demuestra que hay menor prevalencia de enfermedad Celíaca en los pacientes con enfermedad inflamatoria intestinal en relación a la población general, siendo más prevalente en la colitis ulcerosa.

Unique features of primary sclerosing cholangitis in children

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Current Opinion in Gastroenterology 2010, 26:265–268

Purpose of review

To summarize publications on juvenile primary sclerosing cholangitis (PSC) published over the past 5 years. These studies contribute to the understanding of the clinical features, diagnostic pathways, genetic aspects, treatment and outcome of this condition.

Recent findings

Sclerosing cholangitis with strong autoimmune features is particularly frequent in paediatric age, where it is more common in girls, responds to immunosuppressive treatment and has a better prognosis than classical PSC. Modern-day magnetic resonance cholangiography allows accurate diagnosis of bile duct disease in most cases. Prolonged oral vancomycin treatment may be beneficial in difficult-to-treat PSC associated with inflammatory bowel disease. Juvenile PSC has a high recurrence rate after liver transplantation. PSC susceptibility and resistance are associated with both human leucocyte antigen-related and unrelated genetic factors.

Summary

Studies on large cohorts of children with PSC are needed for a better understanding of pathogenic mechanisms, response to treatment and outcome of this serious condition.

Keywords

autoimmune sclerosing cholangitis, inflammatory bowel disease, magnetic resonance cholangiography, primary sclerosing cholangitis

Curr Opin Gastroenterol 26:265–268

0267-1379

CLINICAL REVIEW

Hepatopancreatobiliary Manifestations and Complications Associated with Inflammatory Bowel Disease

Udayakumar Navaneethan, MD, and Bo Shen, MD

IBD/ CEP

- Prevalencia IBD – Colangitis esclerosante= entre el 2 al 7,5%
- 85% Colangitis esclerosantes se asocian a Enfermedad Inflamatoria.
- 1 a 2 % de los pacientes con Enfermedad de Chron presentan Colangitis.
- 5 a 7% pac con C.U presenta CEP
- 19% requiere trasplante hepático dentro de los 15 años de evolución

- IBD + CEP = ↑ incidencia de Displasia colónica.

↑ Colangiocarcinoma

- Tratamiento:
  - UDCA
  - Trasplante hepático