Celiac Disease

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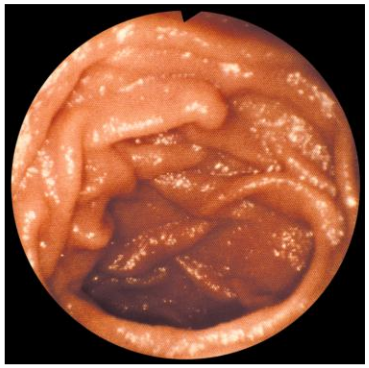
Definition

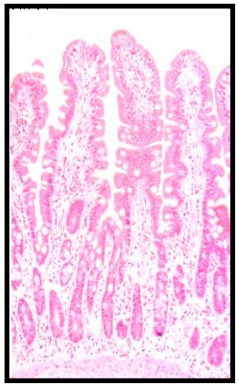
- Celiac disease is an autoimmune condition
- Occurs in genetically susceptible individuals
  - DQ2 and/or DQ8 positive HLA haplotype is necessary but not sufficient
- A *unique* autoimmune disorder because:
  - both the environmental trigger (gluten) and the autoantigen (tissue Transglutaminase) are known
  - elimination of the environmental trigger leads to a complete resolution of the disease

Celiac Disease:

A New Paradigm of Autoimmune Disease

Normal small bowel

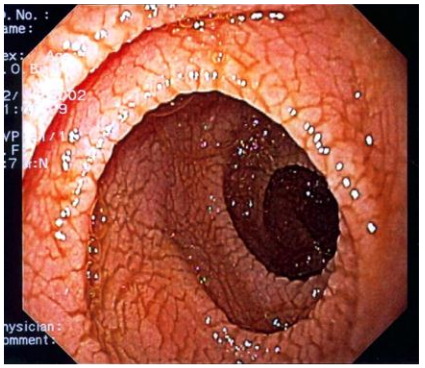


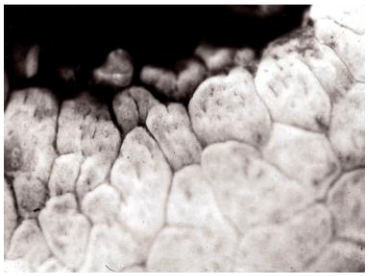


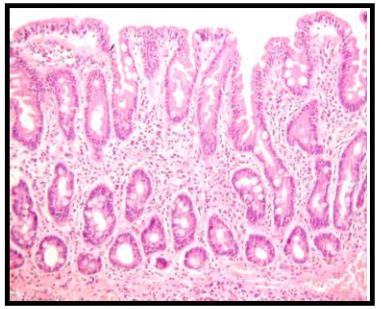
Gluten

Gluten-free diet

Celiac disease

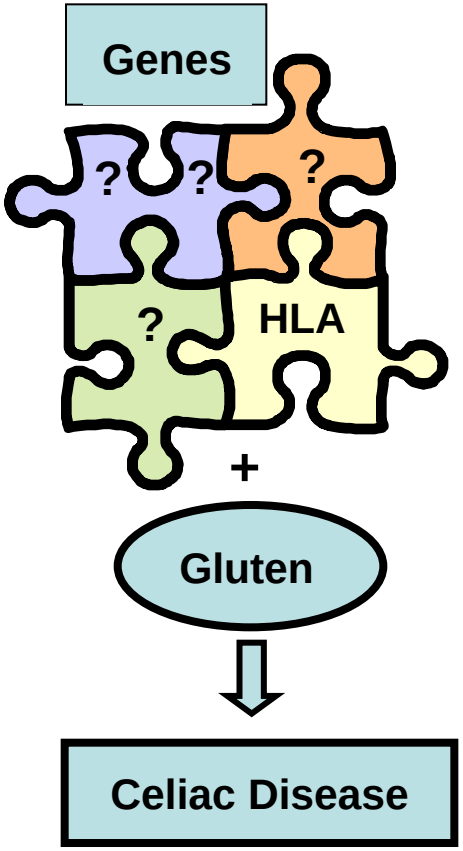






Genetics

- Several genes are involved
- The most consistent genetic component depends on the presence of HLA-DQ (DQ2 and / or DQ8) genes
- Other genes (not yet identified) account for 60 % of the inherited component of the disease
- HLA-DQ2 and / or DQ8 genes are necessary (No DQ2/8, no Celiac Disease!) but not sufficient for the development of the disease



Dietary Factors

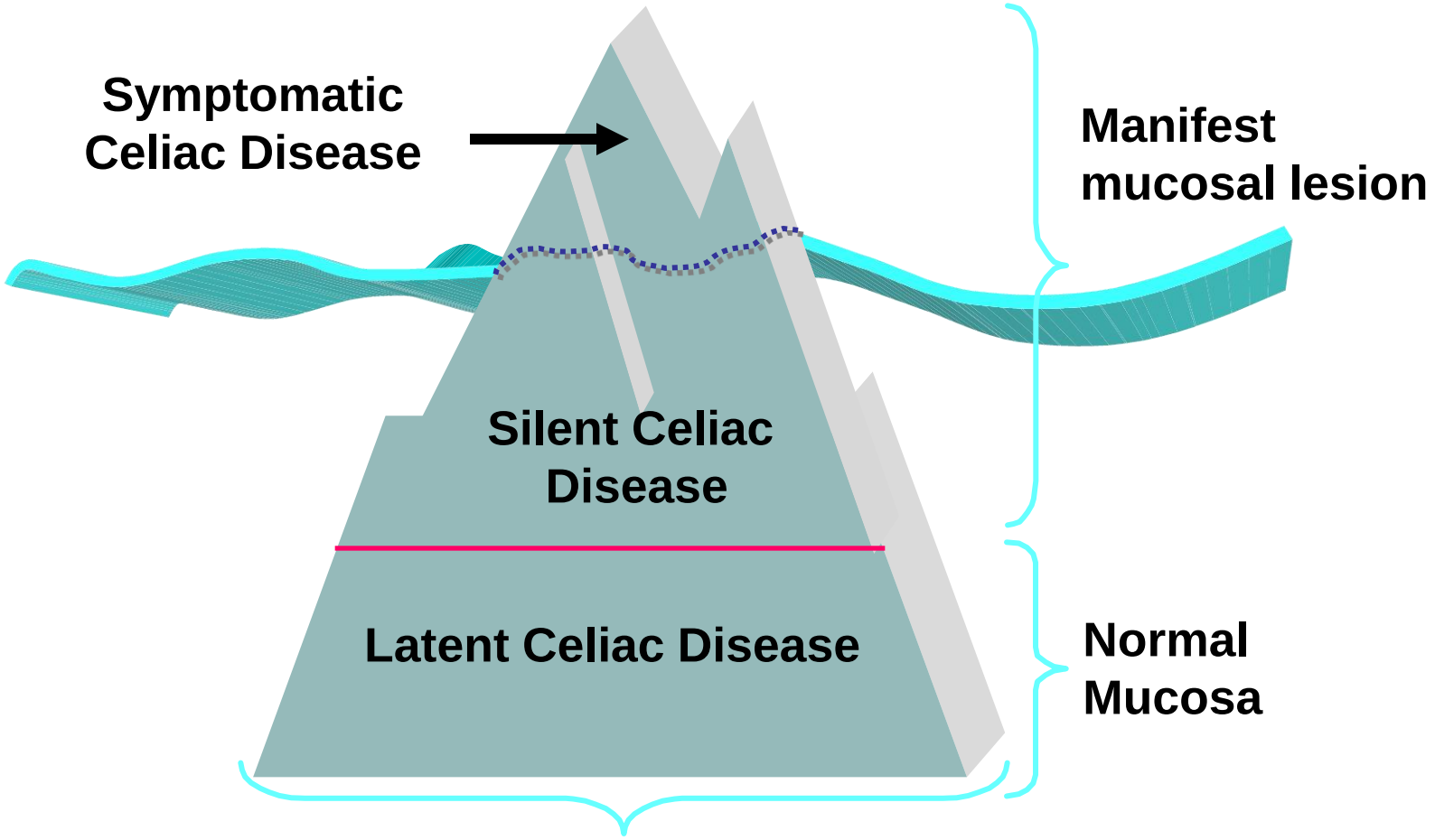
The Grass Family - (GRAMINEAE)

Subfamily — **Festucoideae**

Tribe

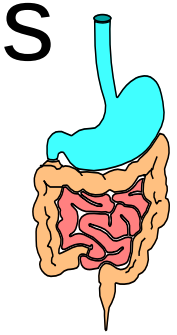
Zizaneae	Oryzeae	Hordeae	Aveneae	Festuceaea	Chlorideae
wild rice	rice	wheat	oat	finger millet (ragi)	teff
		rye			
		barley			

The Celiac Iceberg



Genetic susceptibility: - DQ2, DQ8
Positive serology

Gastrointestinal Manifestations ("Classic")



Most common age of presentation: 6-24 months

- Chronic or recurrent diarrhea
- Abdominal distension
- Anorexia
- Failure to thrive or weight loss
- Abdominal pain
- Vomiting
- Constipation
- Irritability

Rarely: Celiac crisis

IG. 2.—Photograph of five cases of coeliac disease showing the general clinical feature

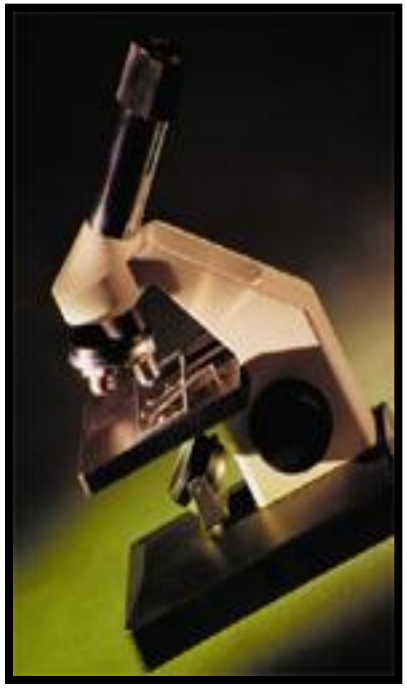
Non Gastrointestinal Manifestations

Most common age of presentation: older child to adult

- **Dermatitis Herpetiformis**
- **Dental enamel hypoplasia of permanent teeth**
- **Osteopenia/Osteoporosis**
- **Short Stature**
- **Delayed Puberty**
- **Iron-deficient anemia resistant to oral Fe**
- **Hepatitis**
- **Arthritis**
- **Epilepsy with occipital calcifications**

Listed in descending order of strength of evidence

Diagnosis



Diagnostic principles

- Confirm diagnosis before treating
  - Diagnosis of Celiac Disease mandates a strict gluten-free diet for life
    - following the diet is not easy
    - QOL implications
- Failure to treat has potential long term adverse health consequences
    - increased morbidity and mortality

Serological Test Comparison

	Sensitivity %	Specificity %
AGA-IgG	69 – 85	73 – 90
AGA-IgA	75 – 90	82 – 95
EMA (IgA)	85 – 98	97 – 100
TTG (IgA)	90 – 98	94 – 97

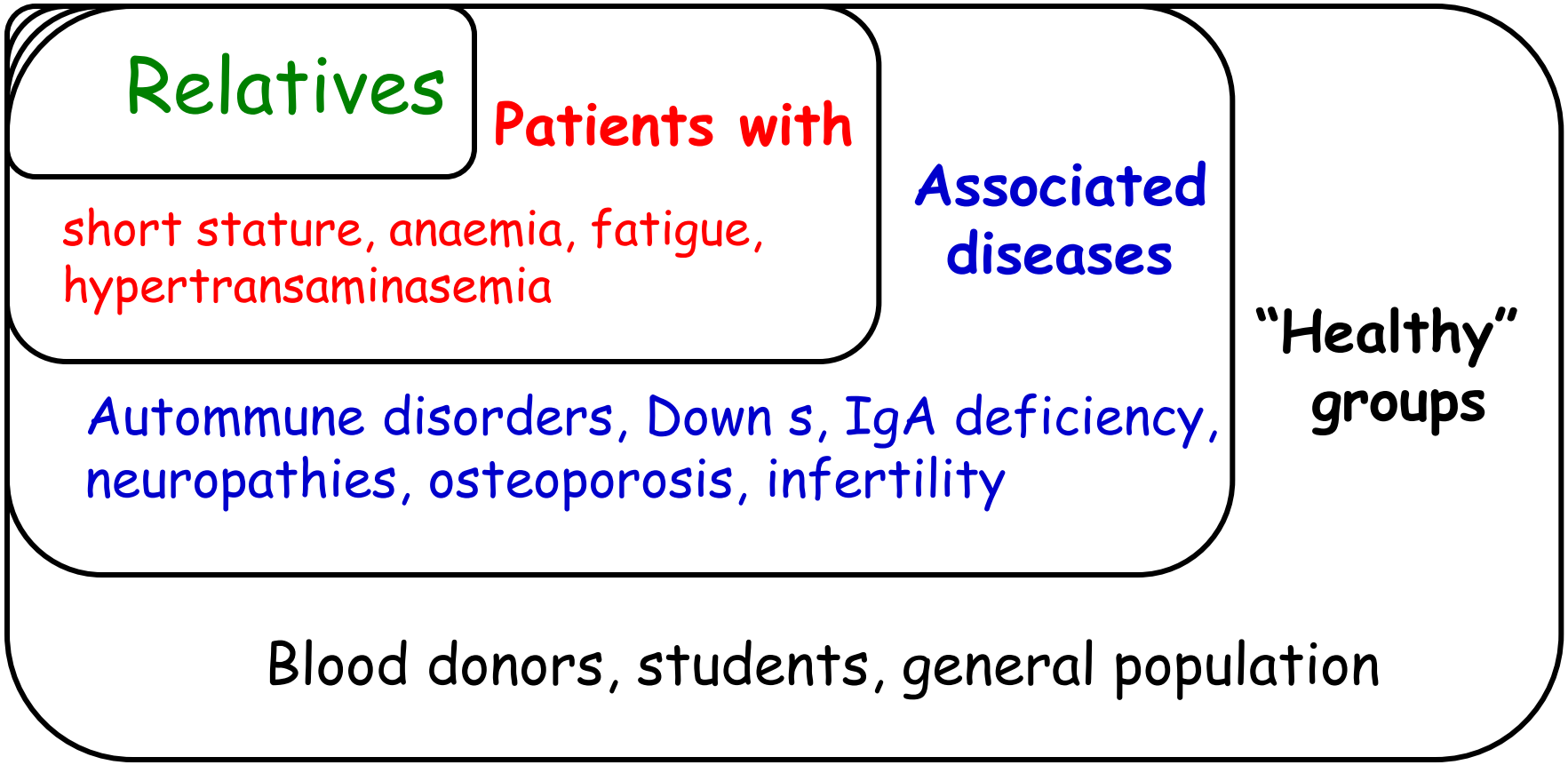
***Farrell RJ, and Kelly CP. Am J Gastroenterol* 2001;96:3237-46.**

Epidemiology

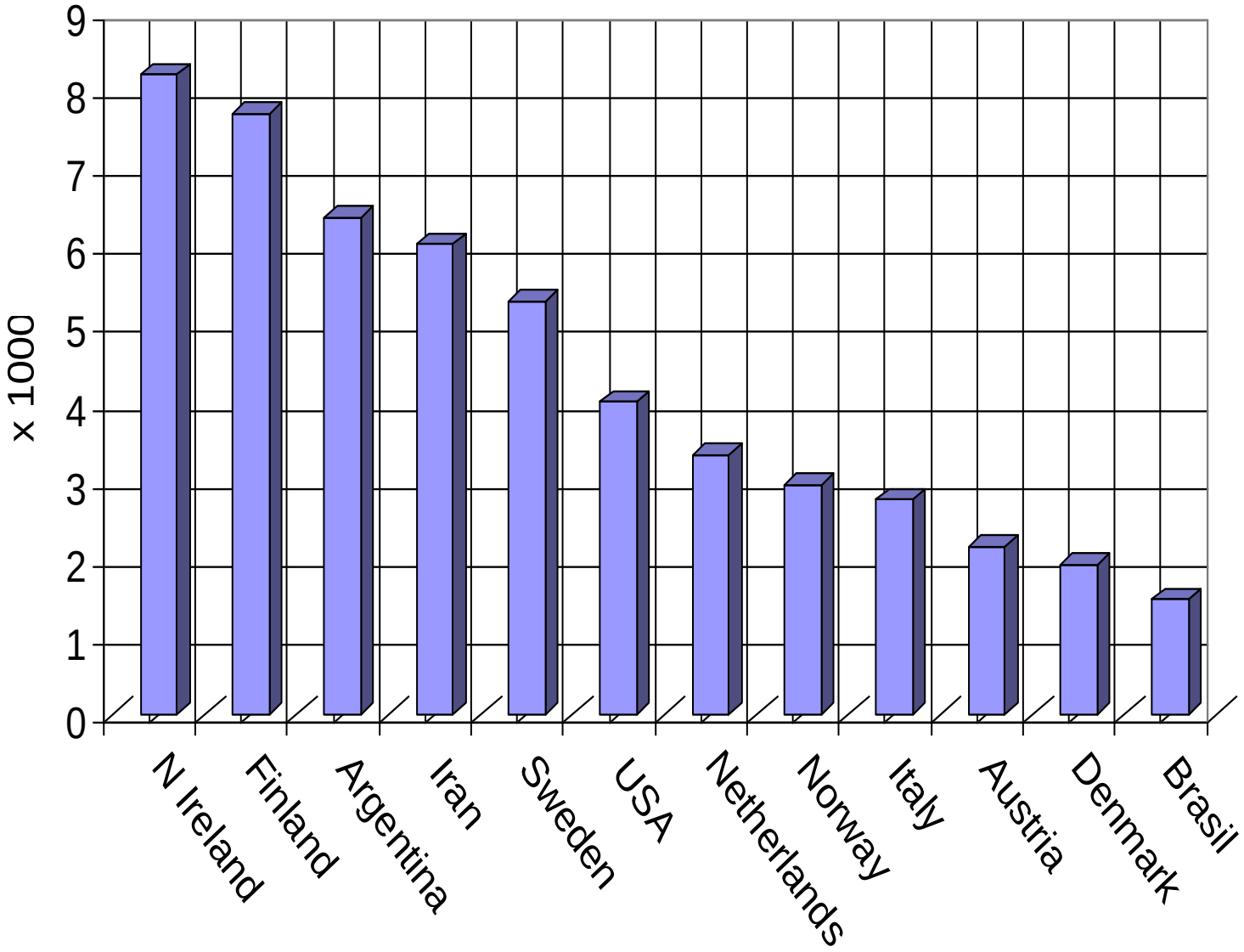
The “old” Celiac Disease Epidemiology:

- **A rare disorder typical of infancy**
- **Wide incidence fluctuates in space (1/400 Ireland to 1/10000 Denmark) and in time**
- **A disease of essentially European origin**

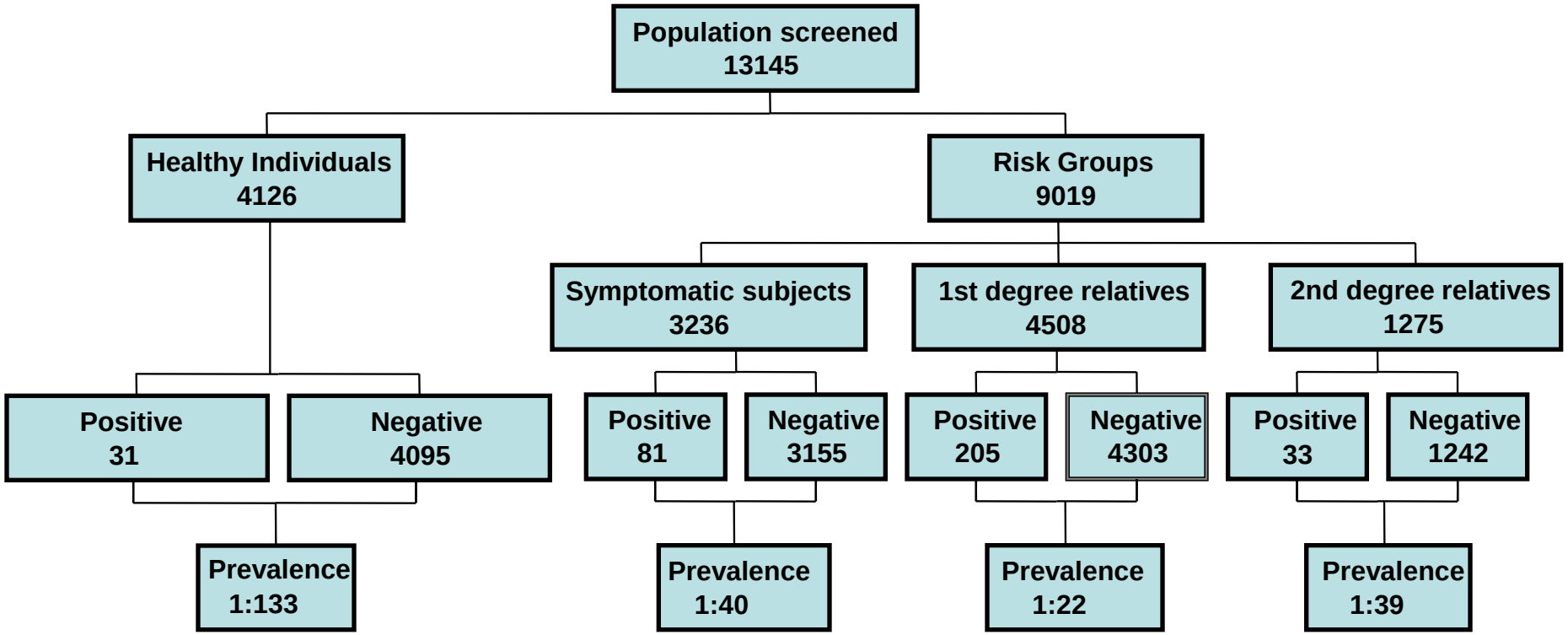
"Mines" of CD have been found among



CD prevalence in blood donors



Celiac Disease Epidemiological Study in USA



Projected number (conservative) of celiac disease patients in the U.S.A.: 2,115,954
MAJOR PUBLIC HEALTH PROBLEM NATIONAWIDE WITH SOME REGIONAL DIFFERENCES

A. Fasano et al., *Arch Int Med* 2003;163:286-292.

Treatment

- Only treatment for celiac disease is a gluten-free diet (GFD)
  - Strict, lifelong diet
  - Avoid:
    - Wheat
    - Rye
    - Barley