# Pediatric Celiac Disease

DATE: August 26, 2020
PRESENTED BY: Mikelle Bassett, MD

# Disclosure

- I disclose that I have no relevant financial relationships with commercial interests related to this content.

# Objectives

- Outline common clinical signs and symptoms of pediatric celiac disease, describe best practice screening and indications for referral to gastroenterologist.
- Describe recommended dietary treatment of celiac disease.
- Identify the psychosocial impact of celiac disease on youth and families, and describe supportive interventions.

# Outline

- Definition
- Epidemiology
- Presentation
- Pathophysiology
- Diagnosis

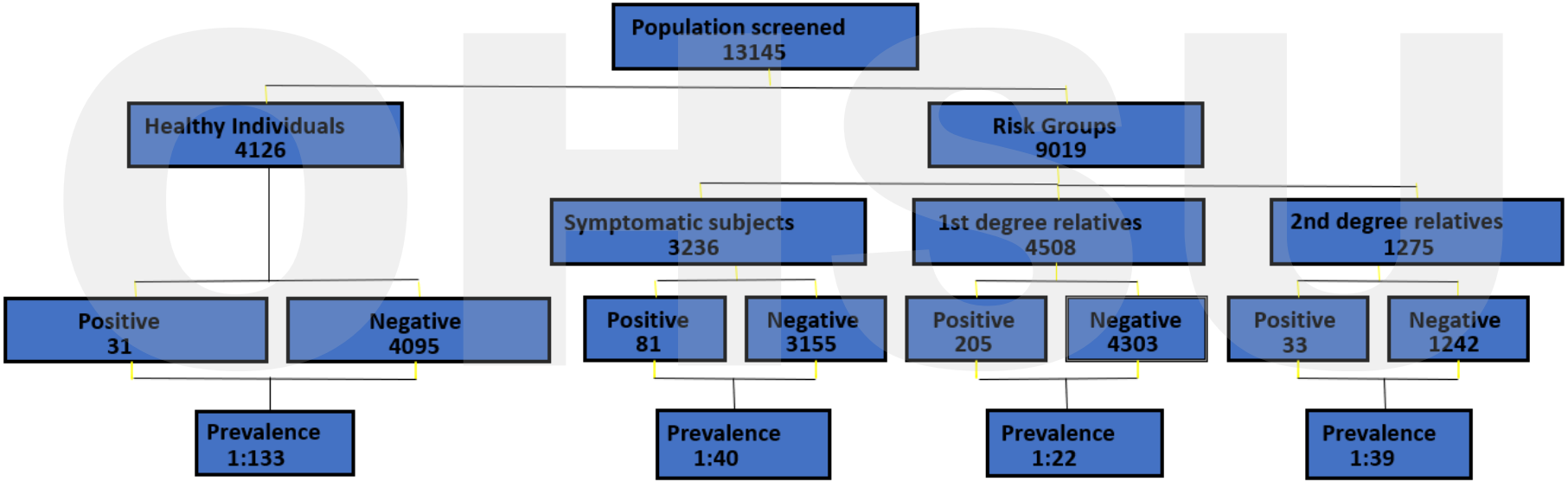
# Definition

- Celiac disease is a complex autoimmune disease, triggered by ingestion of gluten in genetically predisposed individuals.
  - Results in variable degrees of small bowel inflammation
  - Wide range of gastrointestinal and extraintestinal manifestations

# Expanded Definition

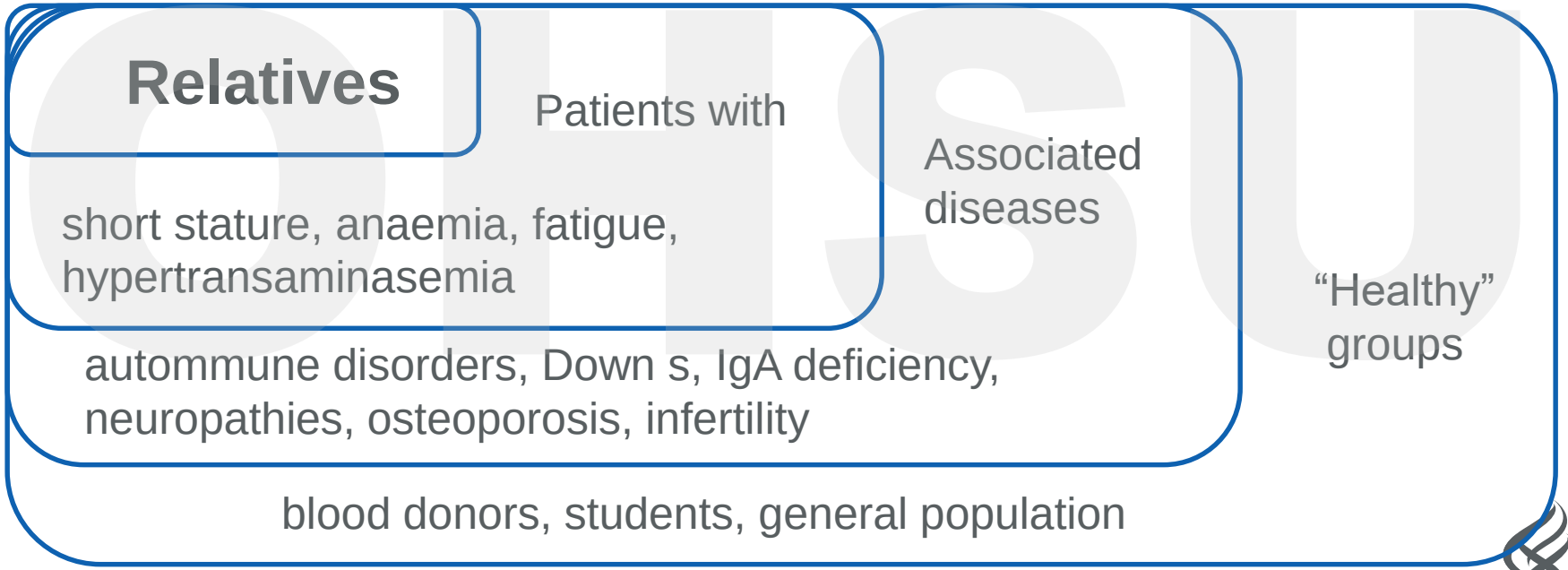
- Celiac disease is an autoimmune condition
- Occurs in genetically susceptible individuals
  - DQ2 and/or DQ8 positive HLA haplotype is necessary but not sufficient
- A unique autoimmune disorder because:
  - both the environmental trigger (gluten) and the autoantigen (tissue Transglutaminase) are known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

# Celiac Disease Epidemiological Study in USA

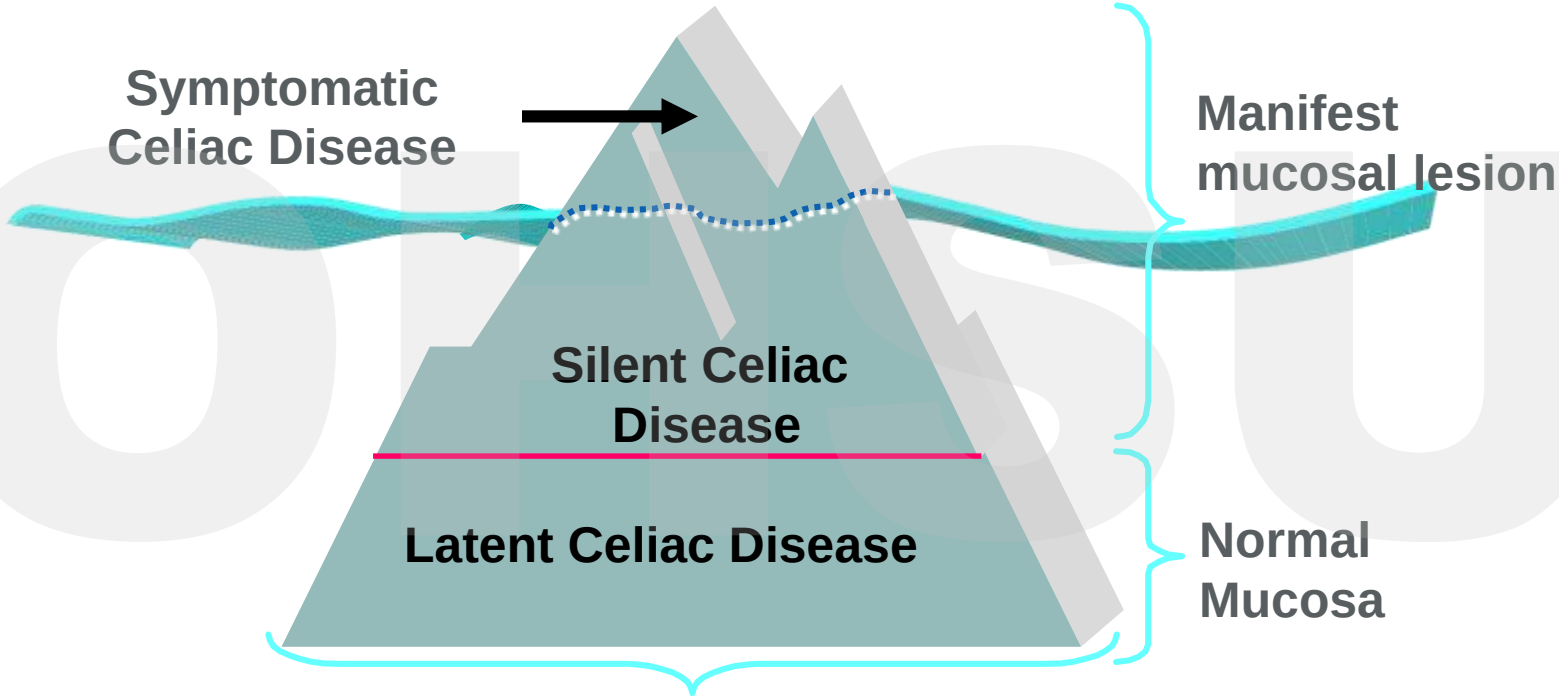



**Projected number of celiacs in the U.S.A.: 2,115,954**
**Actual number of known celiacs in the U.S.A.: 40,000**
**For each known celiac there are 53 undiagnosed patients.**

# “Mines” of Celiac Disease Were Found Among:



# The Celiac Iceberg




Genetic susceptibility: - DQ2, DQ8
Positive serology

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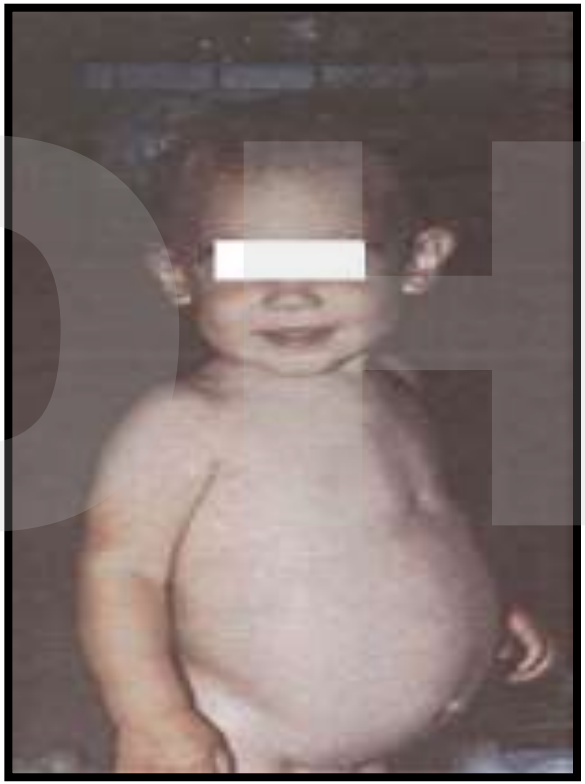
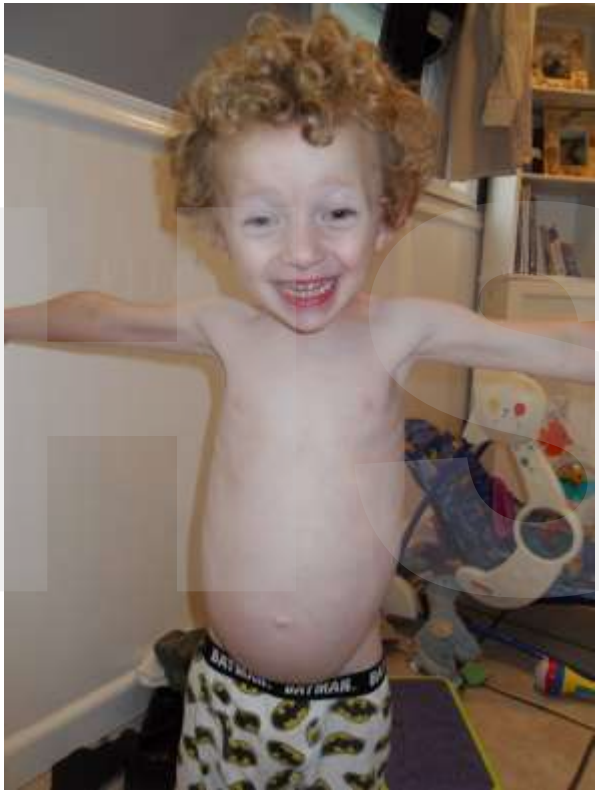
# “Classic” Celiac Presentation

- i.e. Toddler Celiac
- 6-24 months of age
- Chronic diarrhea, anorexia, abdominal pain, FTT, pale stools and/or vomiting.
- Can progress to severe malnutrition-- temporary immunodeficiency can be present as well.





# Childhood Celiac Disease

- **Variable**
- Diarrhea, constipation, bloating, vomiting
- Extraintestinal symptoms more common than in classic/toddler form
- It is another “Great Masquerader”!

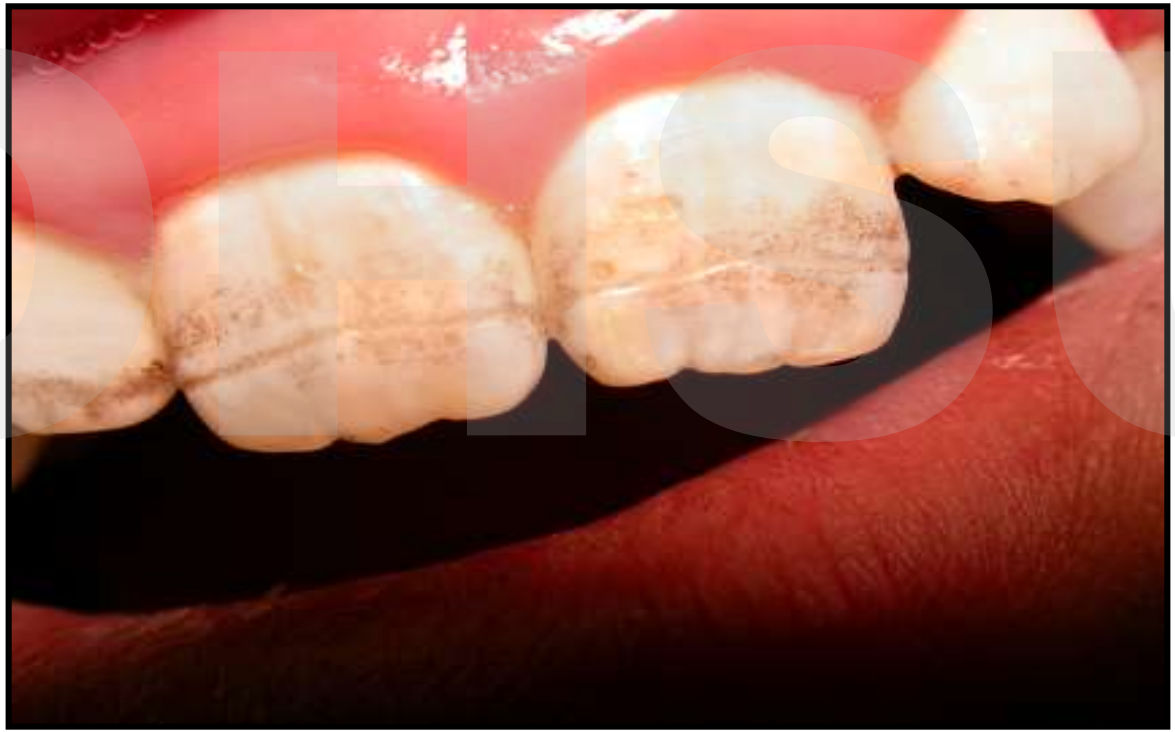
# Non Gastrointestinal Manifestations

- Dermatitis Herpetiformis
- Dental enamel hypoplasia of permanent teeth
- Osteopenia-fracture not explained by level of trauma
- Short Stature
- Delayed Puberty
- Recurrent aphthous stomatitis
- Iron-deficient anemia resistant to oral Fe
- Arthritis or arthralgia
- Elevated transaminases

# Short Stature/Delayed Puberty

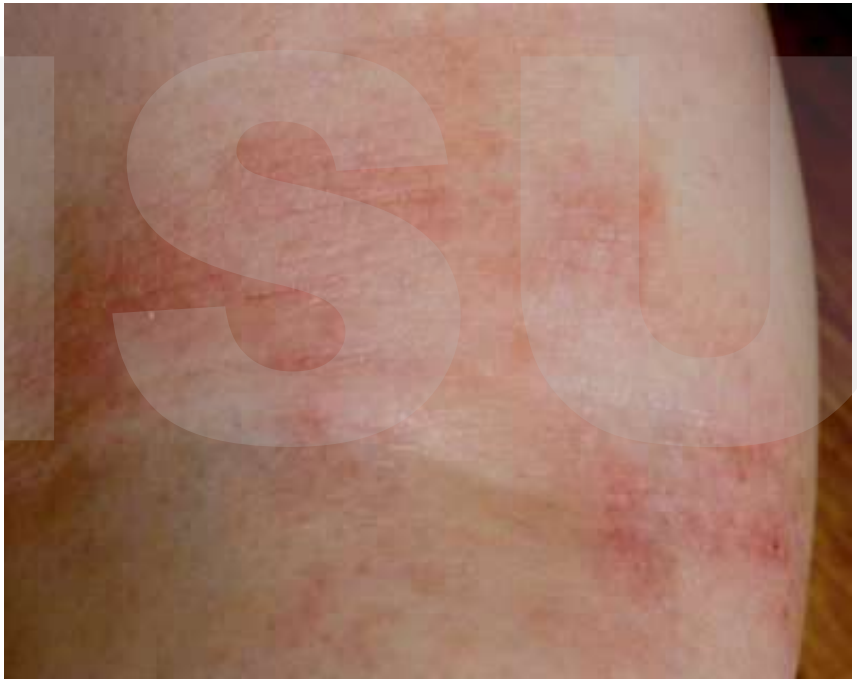
- Short stature in children / teens:
  - ~10% of short children and teens have evidence of celiac disease
- Delayed menarche:
  - Higher prevalence in teens with untreated Celiac Disease

# Dental Enamel Defects



# Skin Disorders associated with celiac disease

- Acquired ichthyosis
- Cutaneous amyloid
- Dermatitis herpetiformis
- Eczema
- Epidermal necrolysis
- Nodular prurigo
- Pityriasis rubra pilara
- Pustular dermatitis



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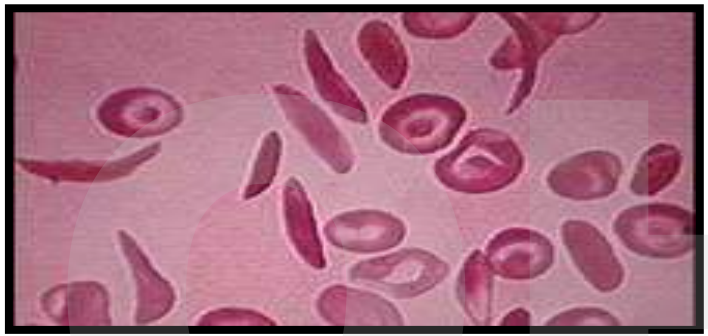
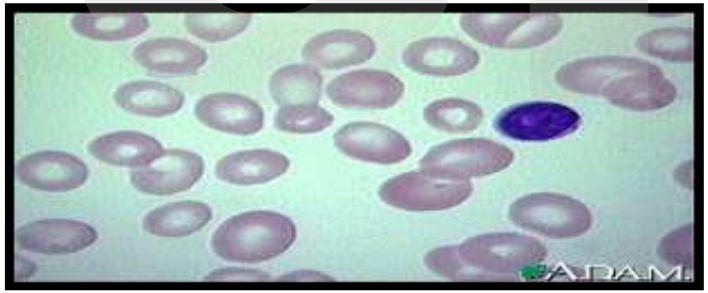
# Fe-Deficient Anemia Resistant to Oral Fe

- Most common non-GI manifestation in some adult studies
- 5-8% of adults with unexplained iron deficiency anemia have Celiac Disease
- In children with newly diagnosed Celiac Disease:
    - Anemia is common
    - Small amount of evidence that Celiac Disease is common in children presenting with anemia

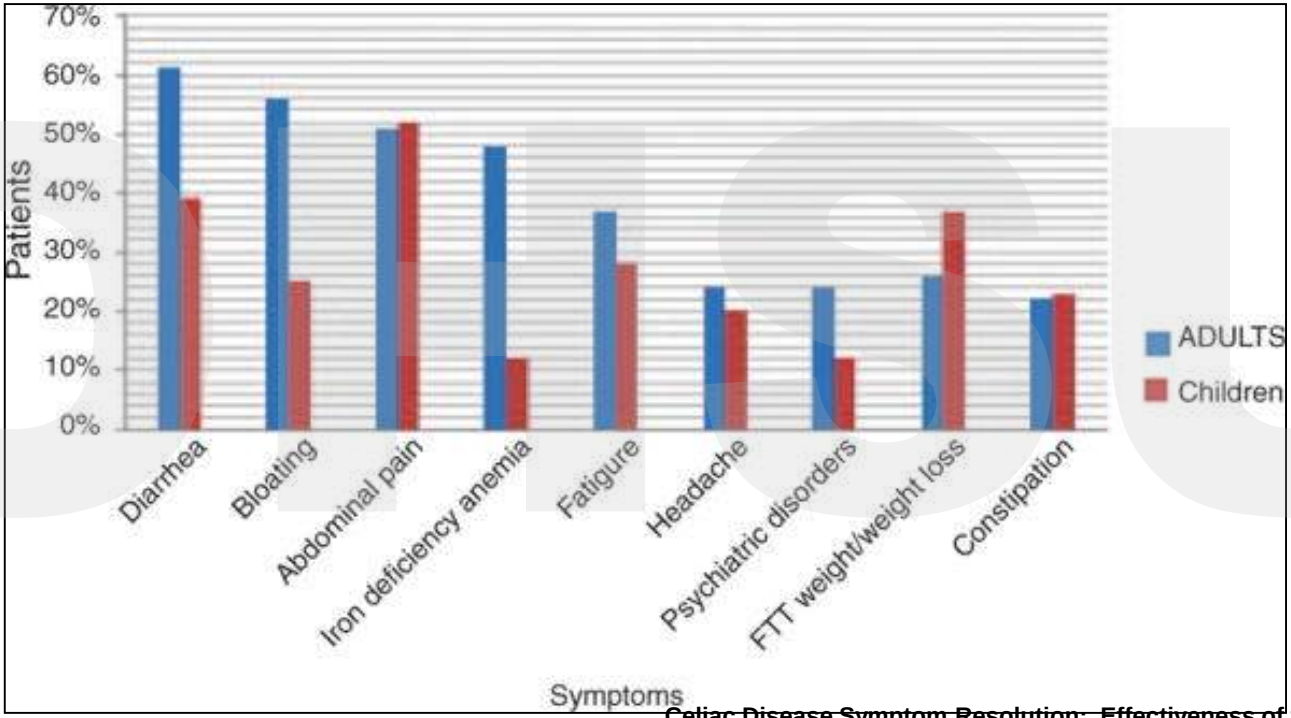
# Anemia in Celiac Disease





- Microcytic anemia - iron absorption most efficient in the duodenum
- Megaloblastic/Macrocytic anemia – folate is absorbed primarily in the proximal third of the small intestine

# Most common gastrointestinal and extra-intestinal manifestations of celiac disease at presentation in adults compared with children.




**Celiac Disease Symptom Resolution: Effectiveness of the Gluten-free Diet.**
Sansotta, Naire; Amirikian, Karine; Guandalini, Stefano; Jericho, Hilary
Journal of Pediatric Gastroenterology & Nutrition. 66(1):48-52, January 2018.
DOI: 10.1097/MPG.0000000000001634

# Range of Sensitivity and Specificity and Use of Current Serologic Tests for Celiac Disease

**Table 3. Range of Sensitivity and Specificity and Use of Current Serologic Tests for Celiac Disease[a]**

| Serologic Study | % Sensitivity | % Specificity | Application in Clinical Practice |
|---|---|---|---|
| IgA tTG | 73.9-100 | 77.8-100 | First-line testing to screen for celiac disease[b] |
| IgG DGP | 80.1-96.9 | 86.0-96.9 | First-line testing for celiac disease in patients with IgA deficiency |
| IgA EMA | 82.6-100 | 94.7-100 | Second-line confirmatory test to screen for celiac disease |
| IgG tTG | 12.6-99.3 | 86.3-100 | Not recommended for routine use because of poor sensitivity compared with IgG DGP |
| IgA DGP | 80.7-95.1 | 86.3-93.1 | Not recommended for routine use because of poor sensitivity and specificity compared with IgA tTG and IgA EMA |

Abbreviations: EMA, antiendomysial antibody; DGP, deamidated gliadin peptide; tTG, tissue transglutaminase.

[a] Adapted from Thawani et al.[41]

[b] Should be sent with a baseline IgA level initially to ensure there is no IgA deficiency.

**Celiac Disease and Nonceliac Gluten Sensitivity A Review**

JAMA. 2017;318(7):647-656. doi:10.1001/jama.2017.9730

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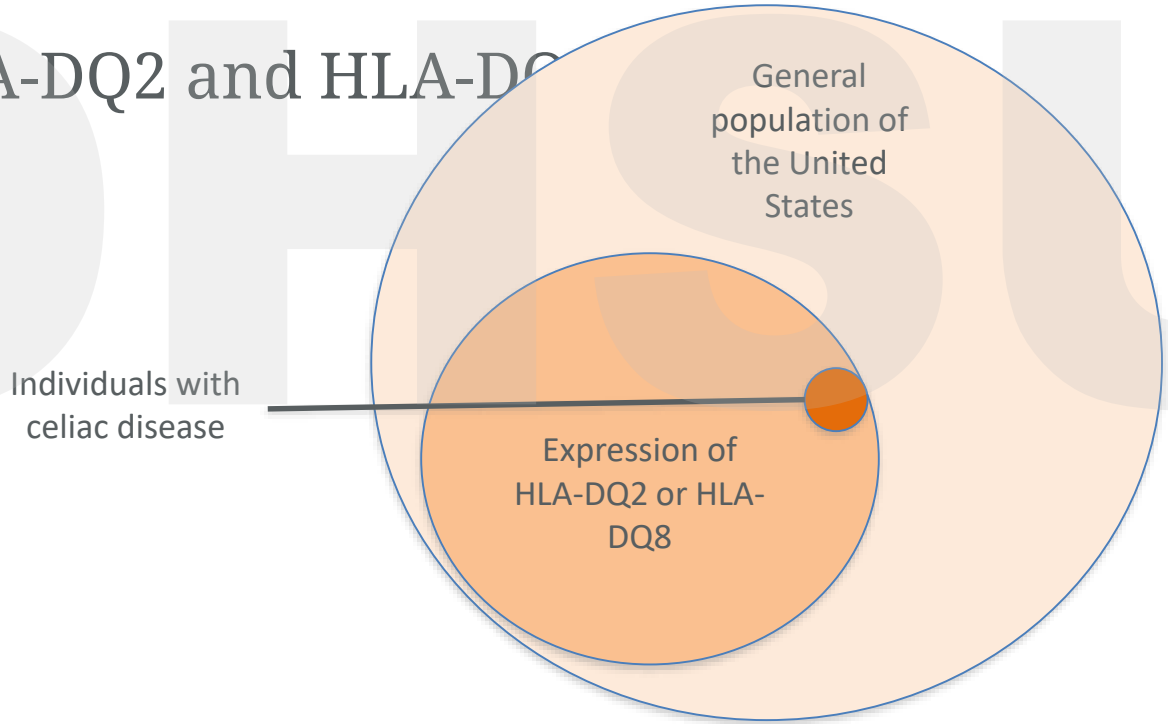
# Testing Pitfalls

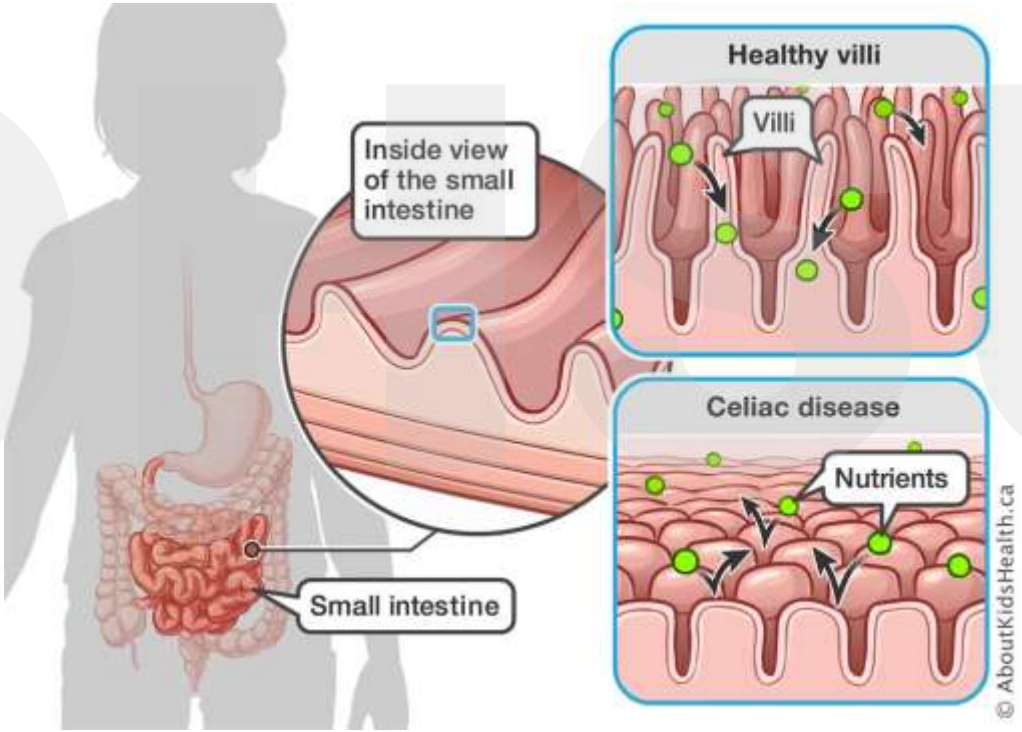
- Under age 2 – Include Deamidated Gliadin Peptide IgG (DGP)!
- Already on a “Gluten Free Diet”—if symptomatic, test
- Different normal TTG IgA range with different labs
- “low” IgA

# Celiac “Genetic Test”

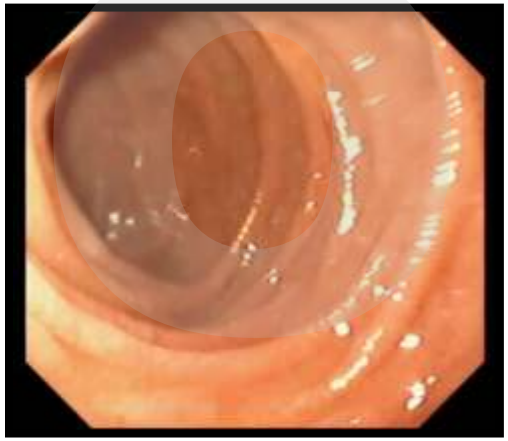
- = HLA-DQ2 and HLA-DQ



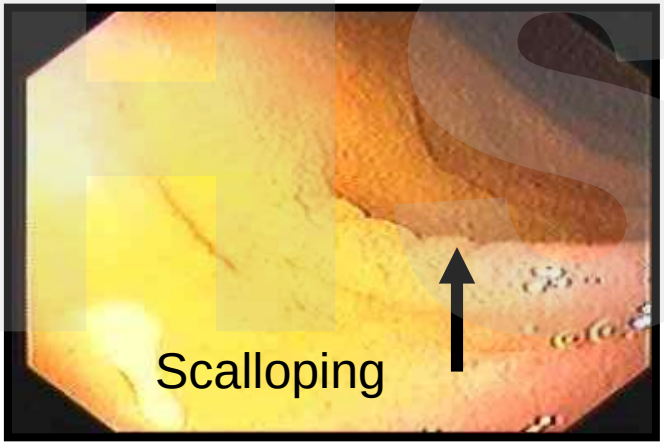
# Diagnosis


Healthy villi
Villi
Inside view of the small intestine
Celiac disease
Nutrients
Small intestine
© AboutKidsHealth.ca

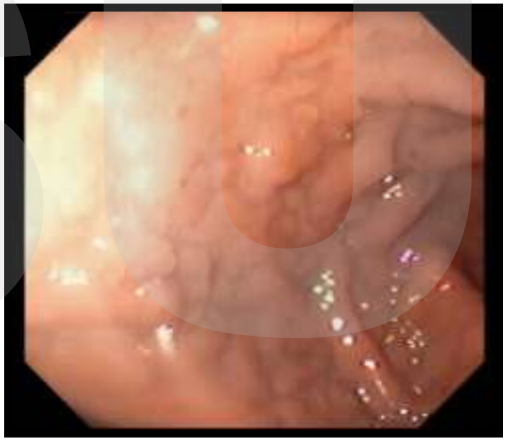
# Endoscopic Findings



Normal Appearing

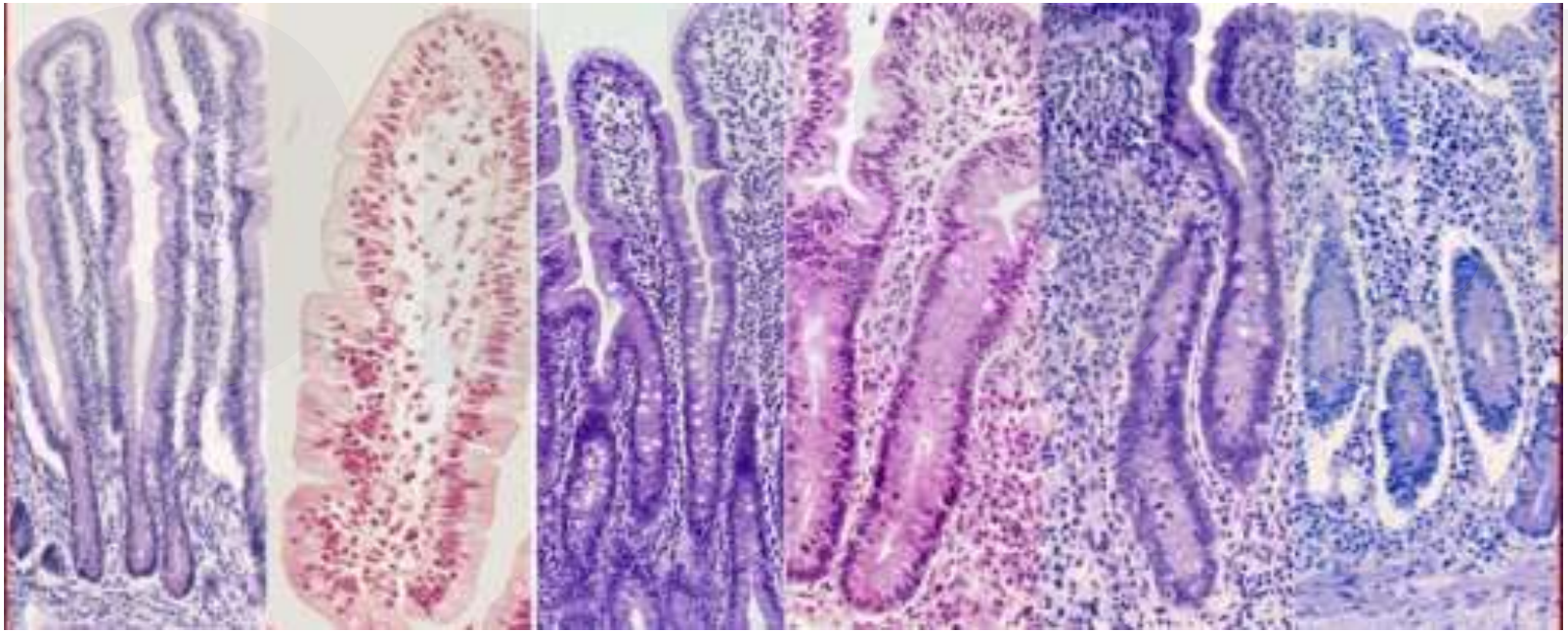



Scalloping



Nodularity

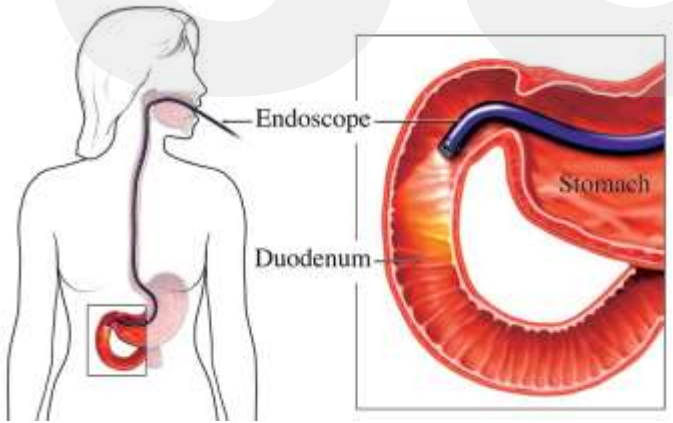
# Marsh Classification



Kneepkens CMP, von Blumberg BME. Clinical practice: Coeliac disease. European Journal of Pediatrics. 2012; 171(7):1011-1021.

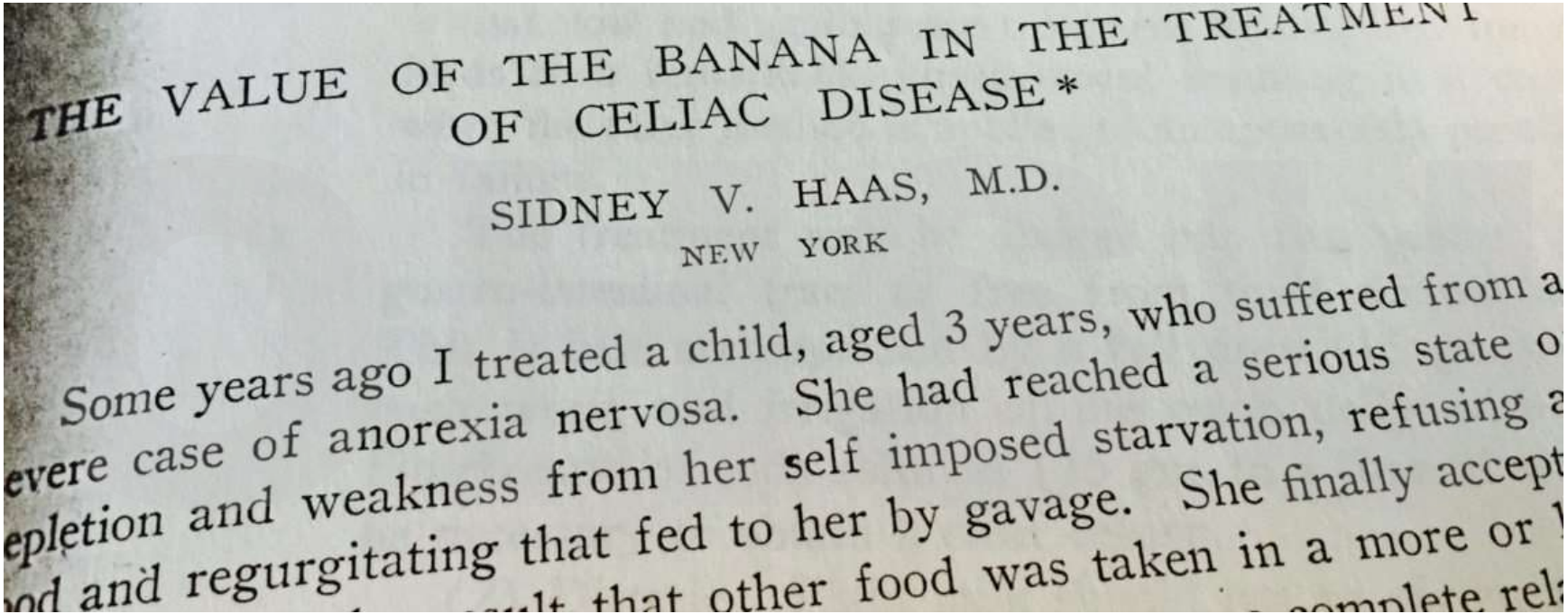
# To Biopsy or Not To Biopsy?

- European guidelines give some criteria to avoid small bowel biopsy, started 2012
- Generally these have been validated
- I'm more hesitant than I used to be about not doing biopsy because later there is often a



# Once Upon a Time….

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



# THE VALUE OF THE BANANA IN THE TREATMENT OF CELIAC DISEASE*

SIDNEY V. HAAS, M.D.
NEW YORK

Some years ago I treated a child, aged 3 years, who suffered from a
evere case of anorexia nervosa. She had reached a serious state o
epletion and weakness from her self imposed starvation, refusing a
od and regurgitating that fed to her by gavage. She finally accept
sult that other food was taken in a more or
complete rel

Haas SV. The Value of the Banana in the Treatment of Celiac Disease. *Am J Dis Child*. 1924; 28(4):421-437.

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and the erythrocytes 5,000,000. This child made very rapid progress, and when last seen, Jan. 12, 1924, his height was 85 cm., his weight 32 pounds 14 ounces (14.5 kg.). The stools were yellow and normal. He walked and was happy and well. His diet is still restricted.

SUMMARY

Celiac disease is a nutritional disturbance of late infancy and early childhood, due to inability to utilize fats and carbohydrates in a normal

*Fig. 10 (Case 7).*—M. R., aged 2 years. Recovery begun.

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# 1942

Bananas Are Her Medicine

A victim of celiac, rare disease that calls for a banana diet, Helene Gottlieb, 15 months, thanks Patrolman Andrew Radtke for finding her a bunch of the now scarce fruit in Brooklyn.

New York, July 21—(AP)—Brooklyn police hunted on foot, in radio cars and by telephone for ripe bananas to aid an ailing child—and came up with 24.

The police had been called in by Mrs. Mitchell-Gottlieb, who said she was unable, because of the banana shortage due to the U-boat campaign against shipping, to feed her child, Helene, 15 months old. The child is suffering from celiac, an infant disease best treated by a diet of the tropical fruit.

After a widespread search yesterday, a policeman on telephone duty located a wholesaler who had eight edible bananas. These were picked up in a radio car and rushed to the Gottlieb home. Sixteen more bananas, which turned up later, also got a ride in a radio car.

About 200 children in the city have celiac, a rare form of dysentery. Last week, 25 pounds of bananas were flown from New York to Montreal to aid celiac-stricken Margo Bradley, 22 months old.

The United Fruit Co. said it would make every effort to supply bananas to celiac cases. When consigned to sufferers of the disease, the company said, bananas will receive priority shipment. A banana powder substitute can be used if necessary, it was said.

The disease is caused by inability to take fats and carbohydrates.

Acta Paediatrica 42: 34—42. Jan. 1953

From the Children's Hospital University of Utrecht (Netherlands), Head: Prof. Dr. A. TEN BOKKEL HUININK, Juliana Children's Hospital, The Hague, Head: Dr. W. K. DICKE and Central Institute for Nutrition Research T. N. O., Utrecht Head: Dr. M. VAN EEKELEN



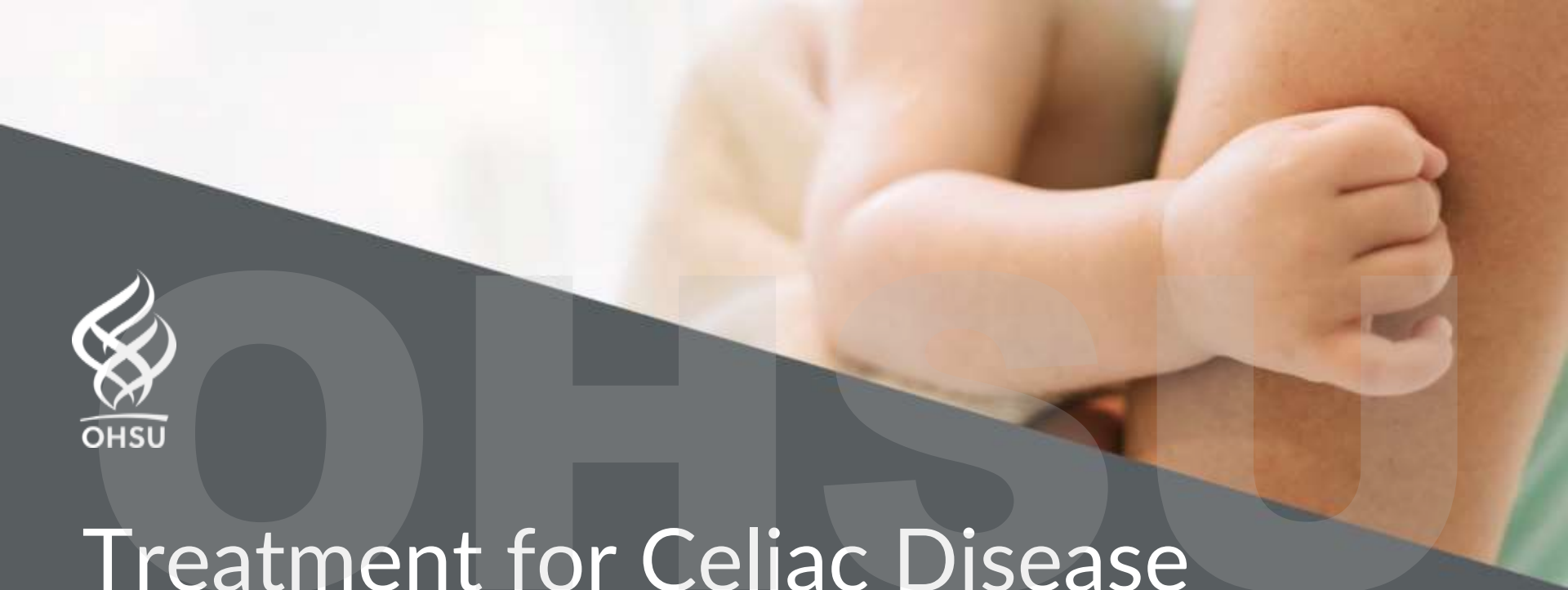
# Coeliac Disease

## II. *The Presence in Wheat of a Factor Having a Deleterious Effect in Cases of Coeliac Disease*

*by* W. K. DICKE, H. A. WEIJERS and J. H. v. D. KAMER

*A basic principle of current opinion on the dietary treatment of c*
*ase is that all starch-containing foodstuffs (with the exception of bar*
*injurious for the patient and must be avoided (*HAAS, ANDERSEN,
*LOWE and others).*
*contra-distinction to this view, we have learnt in the course o*

# Potential Pharmaceutical Treatments..



# Treatment for Celiac Disease

DATE: August 26, 2020

PRESENTED BY: Briza York, RD, CSP, LD, CD
Clinical Pediatric Dietitian
Certified Specialist in Pediatric Nutrition

# Treatment

- Strict adherence to a gluten free diet
- Gluten is food protein found:
  - Wheat: graham, durum, semolina, farro, emmer, spelt, farina, kamut, einkorn
  - Rye
  - Barley: malt and Brewer's yeast
  - Triticale
- Oats need to be specified as gluten free

# Gluten Free Diet Education

- Pt and family meet with registered dietitian for lengthy education
- Focus on foods that can be consumed
- Review foods that need to be avoided
- Review non-food items to check
  - Details depend on age of child
- Importance of avoiding cross contact

# Grains to Eat

- Corn, rice, potato, teff, quinoa, buckwheat, millet, amaranth, sorghum, nut flours, bean flours, tapioca
- Focus on variety

https://ccsearch.creativecommons.org/photos/d8bf7993-db51-476f-9a5a-10b5565af6ca

# Label Reading

- Labeling
  - Certified Gluten Free
  - “Gluten Free”
  - Ingredient list
- Advisory labels are not recommended to use at this time (“may contain”, “made in the same facility as”, etc.)

# Steps to go Gluten Free

Certified GF Gluten-Free

**1**

Remove obvious sources of gluten: bread, pasta, crackers, cereals, and desserts. Start replacing them with easy substitutions: switch from wheat crackers to rice crackers and from wheat cereal to corn cereal. Eat fruits, vegetables, plain milk/cheese/yogurt, meat/poultry/fish without breading, beans, nuts/seeds, and those grain products you have swapped out.

**2**

Start to label read to look for **wheat** (including graham, durum, semolina, farro, emmer, spelt, farina, kamut, and einkorn), **rye**, **barley** (including malt and Brewer's yeast), triticale (wheat and rye hybrid), and **oat** (unless specified gluten free oat). This is a difficult and time consuming step, but will set you up for long-term success.

*Remember: Oats are naturally gluten free, however they are frequently grown in the same field or processed in the same facilities as wheat and are therefore contaminated with gluten. Oats need to be certified gluten free to be safe for people with Celiac Disease to consume.

**3**

Try new gluten free foods! There are many yummy gluten free foods. Try making a quesadilla using a pure corn tortilla instead of a flour tortilla. Figure out which brand of gluten free bread you like best. There are more gluten free grains than gluten containing grains. This can be a fun step because you get to taste new foods. To make it even more fun, you can invite your friends and family to participate with you!

**4**

Continue to sharpen your gluten finding skills and begin to look at non-food items, such as lip balm, gum, toothpaste, and medications. The pharmacy should be able to help if you are on any prescription medications. Over the counter medications are sometimes labeled, but if not, the manufacturer should be able to answer if a medication is gluten free.

**5**

Minimize cross-contact. It's recommended to purchase a new toaster and replace porous surfaces (i.e. wood cutting boards). Plates, cups, utensils, and cookware/bakeware without large scratches do not need to be replaced. Condiments are easy to contaminate. Squeeze bottles are helpful for mayo, mustard, relish, ketchup, and jam/jelly. Peanut butter is a little difficult in a squeeze bottle, so a separate container would be best.

**6**

Get connected! Find gluten free recipes online! Like or follow gluten free organizations (the Gluten Intolerance Group has a Facebook page that posts daily).

**Goal!**

Once you have mastered these steps, you should be on a gluten free diet and on your way to optimal health!

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# Nutrients of Concern

- Wheat is a dietary source of:
  - Carbohydrates, zinc, selenium, thiamin, niacin, riboflavin, folic acid, iron, magnesium, dietary fiber
- If lactose intolerant, may need to limit dairy, which is a source of:
  - Protein, calcium, magnesium, phosphorus, vitamins A, B6, B12, D, riboflavin, pantothenic acid



Groetch et al, 2017 and https://gluten.org/2019/10/17/nutrient-deficiencies/

# Nutrients of Concern

- Losses from malabsorption:
  - Iron, calcium folate, vitamin B12, and fat soluble vitamins (A, D, E, K)
- High rice intake:
  - Arsenic content



https://sites.dartmouth.edu/arsenicandyou/arsenic-and-gluten-free-diets/ and https://gluten.org/2019/10/17/nutrient-deficiencies/

# Other Considerations

- Cumbersome diet
- Expensive
- Anxiety around food
- Feeling of being different
- Social difficulties

# Adjusting & Re-Adjusting to Celiac

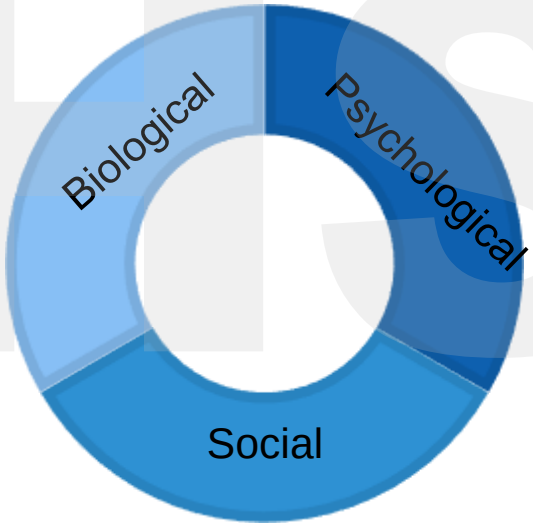
## Psychosocial implications & behavioral health

DATE: August 26, 2020
PRESENTED BY: Jacklyn Stellway, PsyD
Peds GI Psychologist Resident

# Outline

- Celiac disease and mental health
- Developmental stages
- Supporting children across environments
- Adherence & compliance
- Transition
- Referral to behavioral health or child psychology

# Celiac disease and mental health



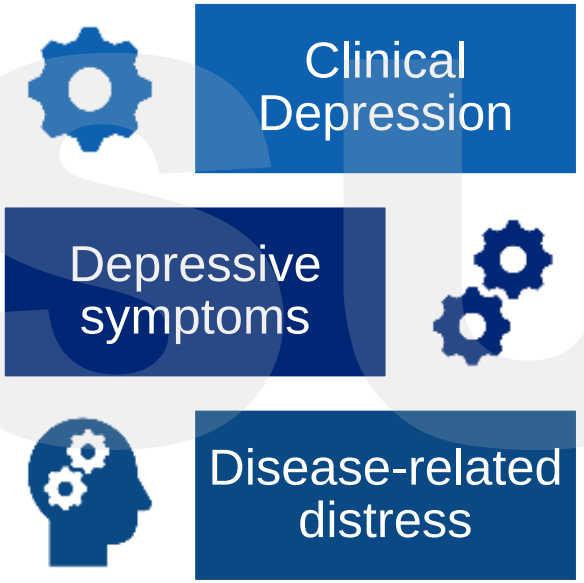
# Emotional distress

- Adjusting to “new normal”
- Symptoms of anxiety, depression, behavioral problems
- Various studies identify increased diagnoses of anxiety, depression, ADHD, compared to healthy controls

- Feelings common with GF lifestyle:
  - Hypervigilance, accidental gluten exposure
  - Anxiety around eating
  - Embarrassed around peers; don’t want to stand out (identified as “different”)
  - Isolated
  - Misunderstood
  - Disappointment, missing out
  - Fatigue from strict adherence
  - Disease burden; burden on family

# Emotional distress

- Measuring depression and anxiety in patients with a chronic illness does not account for CONTEXT
- "Disease-related distress"
  - Expected worries, concerns, fears, and threats that are associated with struggling with a demanding and progressive chronic disease, its management, threats of complications, etc.



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# Developmental Considerations

| Early childhood | Elementary & middle-school age | High-school age | Young adult |
|---|---|---|---|
| Parents are primary source of CD management. | Children are learning self-management, becoming curious, skill-building | Developing independence, autonomy, self-management | More involvement in medical care |
| Age-appropriate learning of one's own health needs | Child & parents manage CD across environments – plan & prepare for activities | Additional skill building – food choices, asking questions, communicating needs | Living independently, making choices, greater responsibility |
| Parents share info and educate other caretakers. | Parents & providers coordinate with school – 504 Plan | Parents & providers support navigation, address non-adherence | Advocate for oneself and navigate social/work environments |



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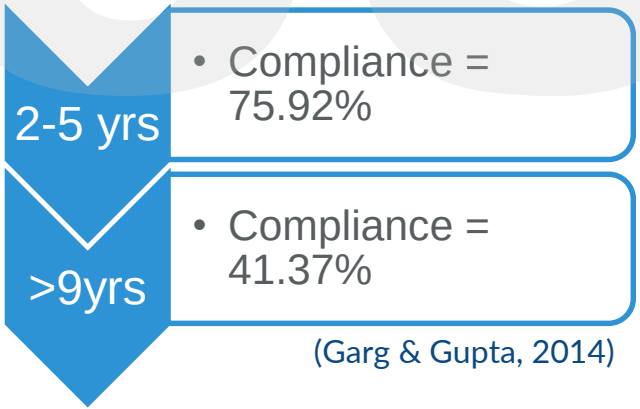
# Celiac Across Environments

- Family – coping, dynamics, finances
  - Parents – guilt, fear, hypervigilance, frustration
- School – birthday parties, holidays
- Relationships with peers and friends – isolation, embarrassment
- Activities outside home – sleepovers, grandparents', camp, extracurriculars


PSYCHOSOCIAL IMPACTS:
FOOD CHOICES, ACCIDENTAL GLUTEN EXPOSURE and BURDEN of the GLUTEN-FREE DIET
JOY BURWELL
ANXIETY
"SOCIO ISOLATION"
GUILT
THE DIFFERENT BURDENS —
CHILDREN — "BEING DIFFERENT"
AT SCHOOL
AT PRIVATE PARTIES
IN COLLEGE TOO
IS IT ABOUT THE DISEASE ITSELF — OR — THE CONVERSATION
HOW DO THEY DEAL WITH IT?
SHARE WITH OTHERS?
KEEP IT TO YOURSELF?
FAMILY TESTING —
HOW DO FAMILY DYNAMICS AFFECT IT?
HOW DOES IT AFFECT YOUR LIFESTYLE?
RELATIONSHIPS
SPOUSES
FRIENDS
SIBLINGS
MOOD DISORDERS
ASPECTS OF DENIAL
JOB — PRODUCTIVITY
— BUSINESS IMPACT
— ECONOMIC IMPACTS
CHALLENGES in EVENT PLANNING
RISK-TAKING — COMPENSATION...
CONSEQUENCES?
"CHEATING"
— ENGAGING IN OTHER RISKY BEHAVIORS
HOW DO YOU COPE?
NOW DRIVING A RACE CAR!
DECISION TO HAVE CHILDREN OR TO ADOPT
WHERE DO YOU GO FOR INFORMATION...
- DOCTOR?
- INTERNET?
HOW COMFORTABLE ARE YOU EXPRESSING YOUR NEEDS?
ATTITUDE SHAPES EXPERIENCE
DEPRIVATION MENTALITY
RATIONING FOOD
BINGING
EATING DISORDERS
PEOPLE'S REACTIONS
IS THIS GLUTEN FREE?
TIME ASPECTS...
DURING THE DAY
HOW OFTEN DO YOU THINK ABOUT FOOD?
TICK TICK TICK TICK
RELIGIOUS ASPECTS —
IMPACT OF FAITH, CHURCHES
IN REALITY IT AFFECTS EVERY ASPECT OF YOUR LIFE!
HOW TO CAPTURE ADOLESCENTS' FEELINGS?
HOW DO YOU PHRASE THE QUESTIONS?
SAME QUESTIONS AS ADULTS?
TWO SETS OF TOOLS
HOW DID YOU REACT WHEN YOU LEARNED OF THE DIAGNOSIS?
"THE FIRST DAY OF A HEALTHY LIFE!"
SUICIDAL THOUGHTS
ETHNICITY
eg. CHINESE
DENIAL
THE IMPORTANCE OF HAVING OPTIONS...
...in RURAL — vs. — URBAN ENVIRONMENTS
FAMILY ATTITUDES, EFFECTS
PARENT
CHILD
THE FAMSS QUALITATIVE TOOL
also FAAMSS
BEYOND CELIAC
awareness • advocacy • action
www.BeyondCeliac.org
— JOE AZAR
crowleyandco.us

# Adherence

- Compliance to GFD varies from 45% to 81% in children (NASPGHN)
- Poor mental health, perception of disease burden, associated with poor compliance
- Statistically significant predictors of compliance: (Garg & Gupta, 2014)
  - Children up to 9 years old
  - Higher level of mother education
  - Nuclear family (vs. joint families)
  - Parents' knowledge and general awareness about the disease




(Garg & Gupta, 2014)

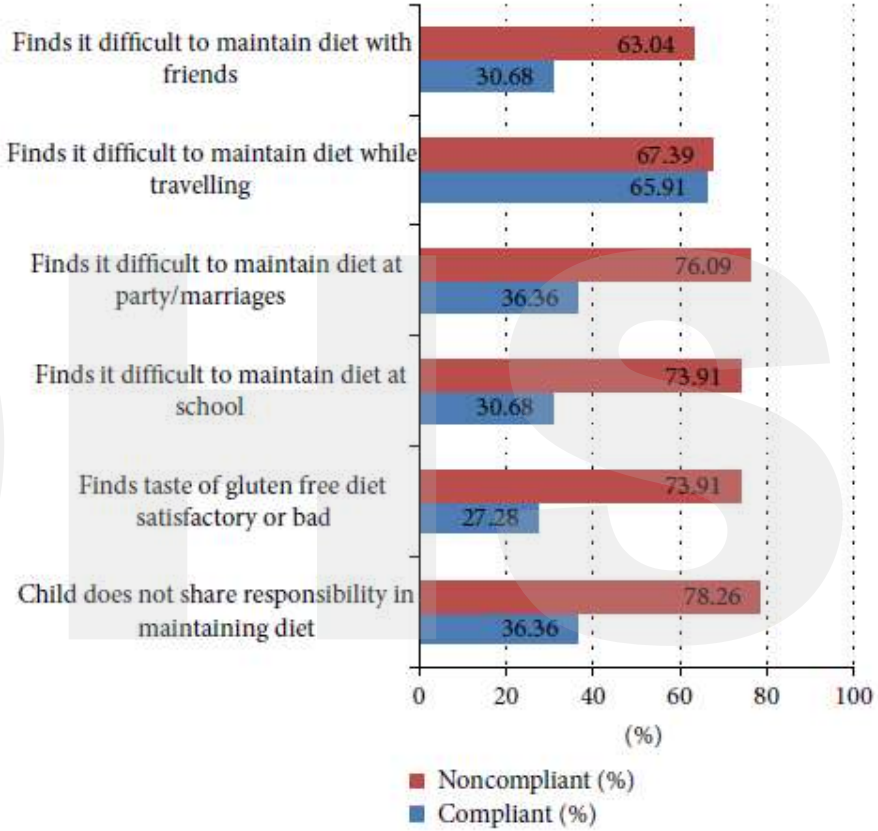



FIGURE 1: Child's attitude towards GFD and its association with compliance.

(Garg & Gupta, 2014)

Table 3: Compliance in relation to parents' attitude.

| Question | Response | Compliant (%) | Noncompliant (%) | $P$ value |
|---|---|---|---|---|
| Finds burden on budget | Heavily | 17 (19.32) | 22 (47.83) | <0.001 |
| | Fairly | 43 (48.86) | 21 (45.65) | |
| | Hardly | 28 (31.82) | 3 (6.52) | |
| Feels burden on self | Y | 18 (20.45) | 25 (54.35) | <0.001 |
| | N | 70 (79.55) | 21 (45.65) | |
| Cooks food once or more than once | >Once | 81 (92.05) | 26 (56.52) | <0.001 |
| | Once | 7 (7.95) | 20 (43.48) | |
| In contact with other parents of children with celiac disease | Y | 30 (34.09) | 6 (13.04) | 0.016 |
| | N | 58 (65.91) | 40 (86.96) | |

(Garg & Gupta, 2014)

# Compliance and age

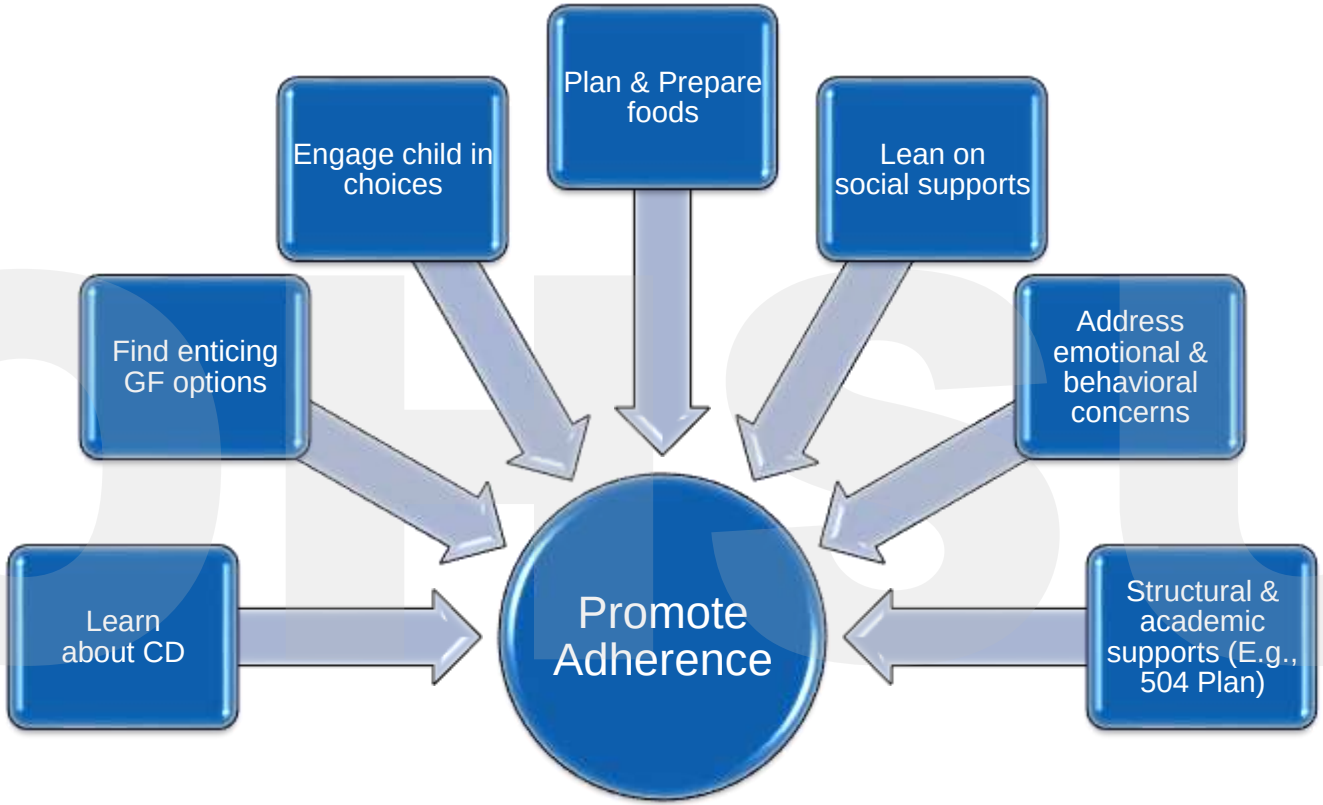
- Adolescents is associated with worse adherence to GFD
- Contributions to non-compliance: (Olsson, et al., 2009)
  - Hectic lifestyle
  - Desire to fit in; be “normal” … not “different”
  - Decreased presence of parents for reminders/guidance
  - Peer pressure
  - Ready-made/packaged foods with unclear labeling
  - Stigma affecting concealment and disclosure; desire to fit in

# “Food that makes you different” - stigma experienced by teens with CD

**Table 2**
**Categories and Subcategories Related to the Stigma Experiences of Swedish Adolescents With Celiac Disease**

| An invisible problem made visible | The center of attention | Facing dietary deviance | In an ideal world |
|---|---|---|---|
| Feelings of social deviance | Knowledge and understanding of others | Confidence in others | To be like everyone else |
| Availability of gluten-free foods | Carelessness of others | Self-protective strategies | Social and practical support |
| Sensory qualities of gluten-free foods | | Concealing—Disclosing | Meeting fellow sufferers |

(Olsson, et al., 2009)




**"***Effective counselling about the diet is the single most important factor to ensure the required restriction in diet***"** (Garg & Gupta, 2014, p. 6)

# GF Adherence Over Time

- Patient-centered, family-focused
- Provider-patient relationship – validation, support
  - Motivational interviewing
- A smooth transition from pediatric to adult care is critical for disease surveillance and treatment adherence (Reilly, et al., 2020)
- Individuals (ages 18-25) who had received a diagnosis of CD before the age of 18:

Belief in the importance of continuing w/ specialist → Successful transition

# Transition to Adult Care

**Do you believe you need to see a specialist because you have celiac disease?**

- 51% “Yes”
- Of those, 46% successfully transitioned to adult care
- Only 14.6% who said “no” successfully transitioned

**Did you receive a referral?**

- 34% “Yes”
- Of those, 50% successfully transitioned to an adult provider
- Only 20% who did not have referral, successfully transitioned

(Reilly, et al., 2020)

# Behavioral health

- When to refer to behavioral health or child psychologist
- Prolonged challenges with adjustment
- Significant anxiety, isolation, sadness, negative thoughts/statements withdrawal (friends and activities)
  - Impacting daily functioning & enjoyment
- Conflicts within relationships (parent-child communication, problem-solving)

# Thank You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# Questions?

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