Interreg
CENTRAL EUROPE

European Union
European Regional
Development Fund

Focus IN CD

# Focus on Coeliac Disease

## Patient centred coeliac disease management

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# About the Focus IN CD project

## Innovative patient centred health care services – advantages of establishing a close CE network in coeliac disease patient health care.

Coeliac disease is a lifelong systemic autoimmune disorder, requiring extremely meticulous dietary treatment. It affects 1-3% of population (up to 5 million in CE) of all ages.

80 % percent of people with coeliac disease remain undiagnosed or misdiagnosed, and diagnostic delays reach 10 years in many regions. Undiagnosed or untreated disease is associated with a number of severe complications and comorbidities.

The Focus IN CD project with full name "Innovative patient centred health care services - advantages of establishing a close CE network in coeliac disease patient health care" addresses specific issues of coeliac disease management. It focuses on the innovative potential in the public sector, better integration of social innovations into the healthcare system with the objective to bring the system closer to the patient, establishing an integrated treatment and ensuring healthy and active ageing of the population. Focus IN CD project is supported by the Interreg Central Europe Programme, funded by the European regional development Fund. A total budget of the project is approximately 1,900,000€. The implementation started in June 2016 and will end in May 2019. Project partners are Municipality of Maribor, University Medical Centre Maribor, E-institute - Institute for Comprehensive Development Solutions, Klinikum der Ludwig-Maximilians-Universitat Munchen, Heim Pal Hospital Budapest, Universita degli Studi di Trieste, Instituto Burlo Garafolo Trieste, Klinički bolnički centar Rijeka, Udruga oboljelih od celijakije Primorsko – goranske županije Rijeka, Coeliac Association from Budapest, Stiftung Kindergesundheit and Primorsko – Goranska županija.

Within the project, we will develop E-tools for healthcare professionals, implement pilot testing on 10 new services and prepare an integrated management model for coeliac disease patients, which can also be applied in other environments, and for other chronic diseases.

Key outputs are Assessment of Coeliac Disease management practices; three developed and implemented E-tools: E-learning tool for HCPs for up-to-date management of CD patients, E-learning tool for patients for necessary every-day life of CD patients and ICT App for HCPs to help them in everyday practice. Ten pilot actions will be implemented and tested to improve skills and competences of health care professionals. The project will demonstrate development and pilot testing of an innovative healthcare service model in management of coeliac disease (CD), which will enable us to develop a patient centred health care service. "Policy recommendations" based on patient centred CD management model will include guidance on how to effectively address the challenges of CD management in the framework of existing healthcare systems.

The public sector is a hidden source of variety of innovation potential. Early prevention of chronic diseases that have severe impact on general well-being of patients is extremely important if we want to reach this goal and ensure sustainable health care systems in CE.

# Coeliac Disease

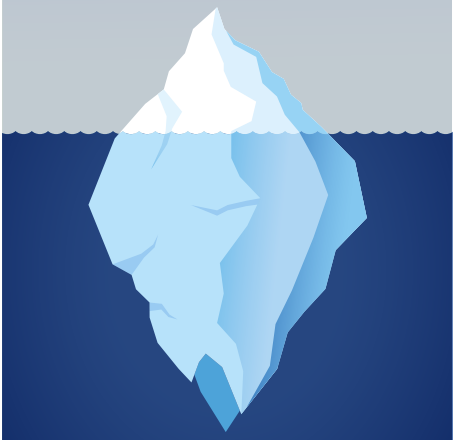
**Coeliac disease is an autoimmune systemic disorder caused by the ingestion of gluten and related proteins, found in wheat, rye, barley and in some cases also in oats in genetically predisposed individuals. It is one of the most common chronic diseases among children and adults and affects about 1% of the population in Europe. Many of patients remain undiagnosed.**

Coeliac disease is a complex disorder strongly associated with HLA DQ2 or DQ8 haplotypes and specific immunological and environmental factors. In coeliac disease patients, ingestion of gluten triggers a chronic damage of the small intestine. A consequence of the morphological changes in the intestinal lining is a weakened function with symptoms of malabsorption. Clinical symptoms characteristic of the disease, such as diarrhoea and malabsorption syndrome, are not the most common forms of the disease anymore. Atypical symptoms and silent forms of the disease are becoming more and more frequent. Based on the clinical picture, coeliac patients can be divided into the following two groups: symptomatic and asymptomatic coeliac disease. The symptomatic coeliac disease can present with gastrointestinal or extraintestinal symptoms and signs. The term asymptomatic or silent coeliac disease is used to refer to patients who were diagnosed with changes characteristic for coeliac disease, although they seem to be asymptomatic.

Diagnosis of coeliac disease is primarily based on the clinical picture. However, the final diagnosis is always based on the presence of a specific reversible immune response and in majority of patients also on detecting histological changes of the small intestine in genetically predisposed individuals. In some cases, the diagnosis can be made without intestinal biopsy. It is important, that patients do not start with a gluten free diet before they receive the final diagnosis.

The only possible way to treat coeliac disease is a very strict lifelong gluten free diet, which improves the clinical picture, normalises the level of antibodies and restores the damaged intestinal lining. Following a strict diet is also the only way to prevent the development of serious long-term effects of the disease. The most significant risk factor for long-term complications is inadequate gluten free diet compliance.

*The coeliac iceberg is large. Representing 1% of total population. However, only a small proportion of these patients is detected, corresponding to the tip of the iceberg. Various data show that only 10% of patients are detected due to symptoms and signs, whereas 90% can remain undiagnosed for a longer period.*



*The size of the submerged part depends very much on patients' awareness and knowledge of HCPs and availability of reliable diagnostic tools.*

# Patients' stories

## We were happy that the marathon from one doctor to another had finished

*The day, when our oldest daughter was diagnosed with coeliac disease was one of the happiest days for our family. The diagnosis "coeliac disease" was among the suspicions of doctors for the best and the least harmful disease. We were happy, that the marathon from one doctor to another had finished and that we finally identified what was wrong and how we can help our daughter. To live and develop into a healthy and happy woman. Our daughter's health problems did not develop overnight, as in a rapid deterioration of her health condition. The changes were very gradual, but still not unnoticed. From the early age of two she had weakened immunity (a hypogammaglobulinaemia) and was more prone to infections, this is why she was managed by specialists in an allergy outpatient clinic. To avoid diseases, she did not attend organised care (kindergarten). Somehow, our happy, but quiet girl, became even more tired and without appetite after the treatment of her last infection. After the consultation with her paediatrician, we did a blood count check, which was fine. Problems with diarrhoea, malaise, pain, constipation or vomiting were absent. I visited the paediatrician`s office several times a month with my moody and tired daughter. No one thought of coeliac disease, we were not referred to a specialist – a gastroenterologist.*

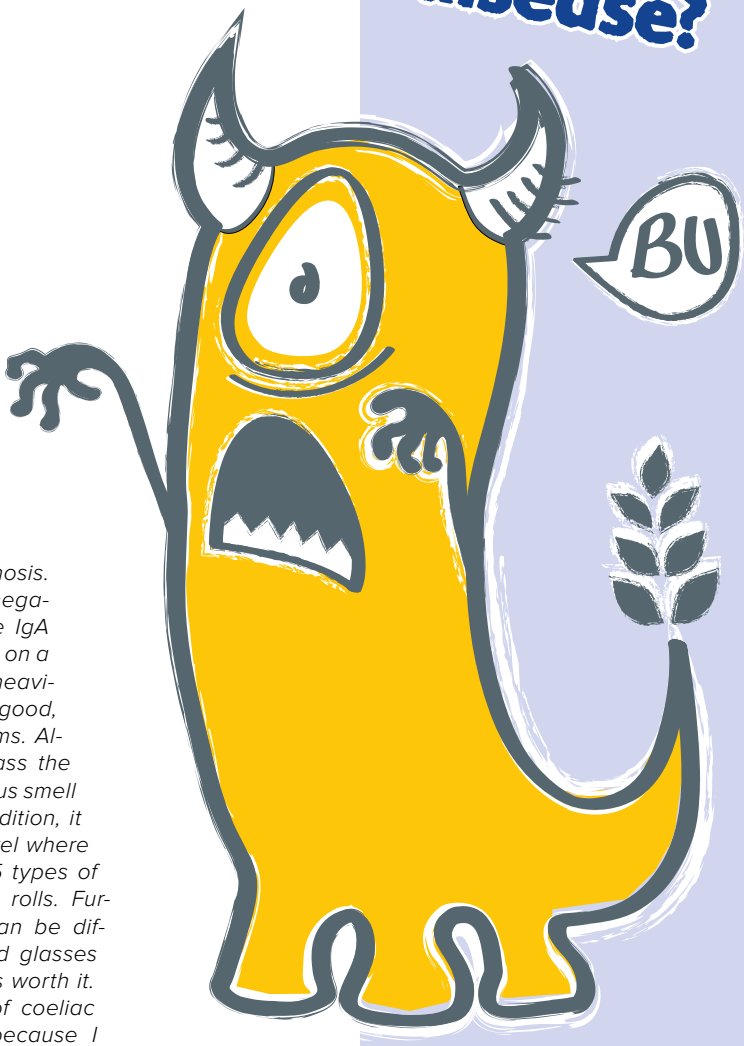
*As we were blessed with a new family member, we thought that maybe it was the lack of acceptance of her sibling and also visited a psychologist. During the holidays we were hoping for an improvement of her health condition, however problems with rapid weight loss and her general condition was very bad. Fortunately we soon had an appointment at the allergist, where we were referred to the gastroenterology department. The diagnosis of coeliac disease was confirmed in 10 days. Our girl, a patient with coeliac disease on a strict gluten-free diet, is growing up and is thriving into a healthy and happy girl. Soon her sister and father, also coeliac patients, joined her in the gluten-free diet.*

Nuša, 11 years, and mom Simona

## The condition was staring me in the face, I just didn't see it

*I was diagnosed with coeliac disease at the age of 44. For many years I had numerous symptoms typical of coeliac disease. From unbearable abdominal pain, diarrhoea, constant bloating, anaemia, fatigue and infections all the time. Since I have thirty years insulin-dependent diabetes, this diagnosis of coeliac disease should be slightly more expected. The condition was staring me in the face, I just didn't see it. As I am a paediatrician who knows about the symptoms, I should have got it sorted sooner. In addition, I know some eminent experts, who are treating the disease in their daily work. A few years earlier, I was traveling to a gastroenterological congress with a colleague expert in the field of coeliac disease. At the time, I suffered severe pain, cramping, bloating, and diarrhoea - especially when I had eaten a good breakfast of fresh rolls, but we didn't see the obvious. In defence of my expert*

# Coeliac disease?

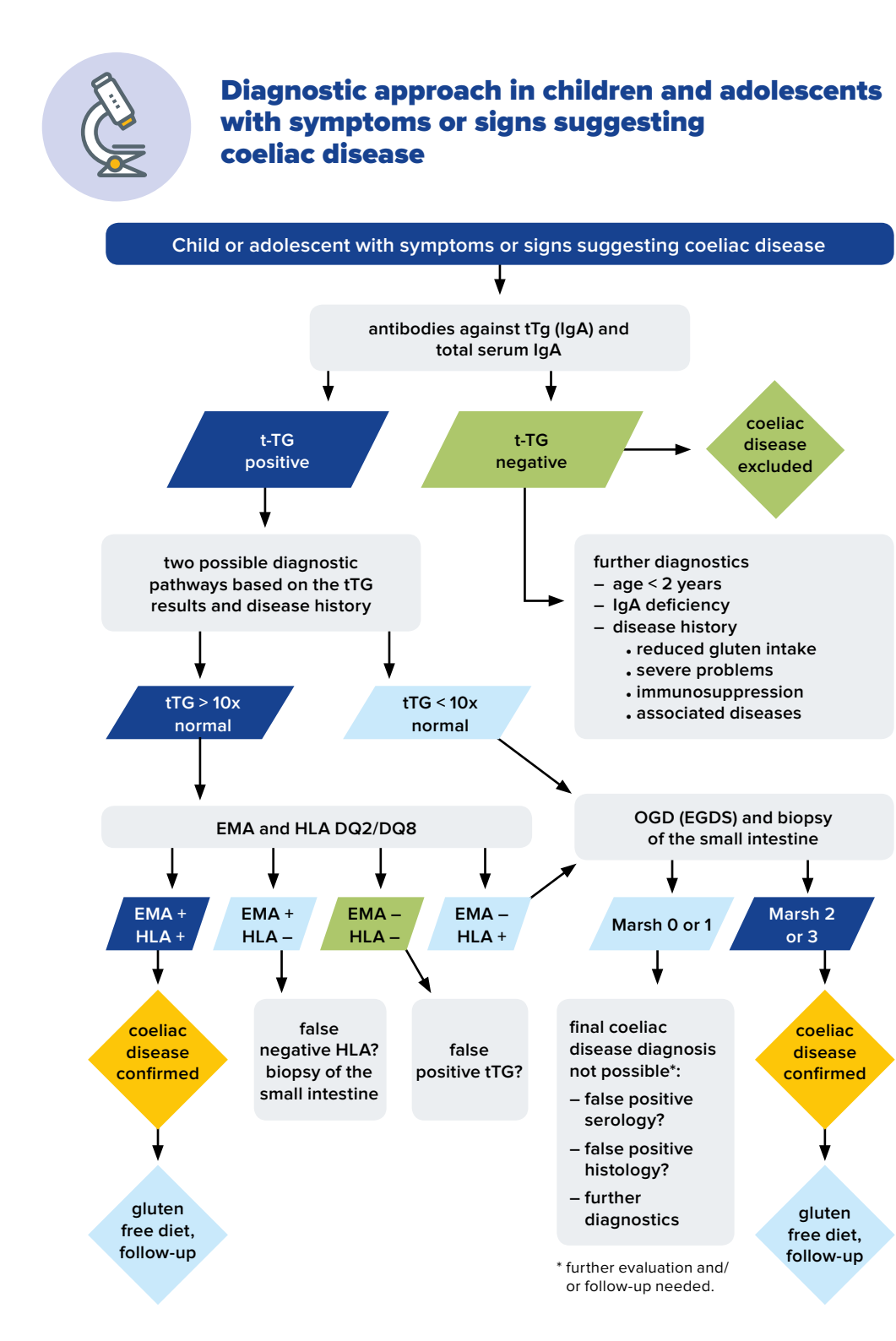



*friend, my coeliac test had been repeatedly reported negative. Afterwards came additional problems. Both my ankles were swollen, my anaemia was severe and iron supplements did not help. Then another colleague saved me and made the diagnosis. My serological test were negative before, because I have IgA deficiency as well. Now, I am on a gluten-free diet, I am 15 kg heavier than when I started. I feel good, without any medical problems. Although it is hard when I pass the bakery and there is a delicious smell of fresh baked bread. In addition, it is hard when I am in the hotel where they have breakfast with 15 types of delicious bread and bread rolls. Furthermore, reading labels can be difficult, especially if you need glasses for small print. However, it is worth it. I accepted the diagnosis of coeliac disease relatively easily, because I am now accustomed to my chronic condition. This is also because my health is incomparable to that of five years ago. I bake my own gluten free bread. I miss some donuts for carnival and Bled cream cake. Nowadays, there is a better variety of gluten-free products. Unfortunately gluten-free products are relatively expensive, which can be a big problem.*

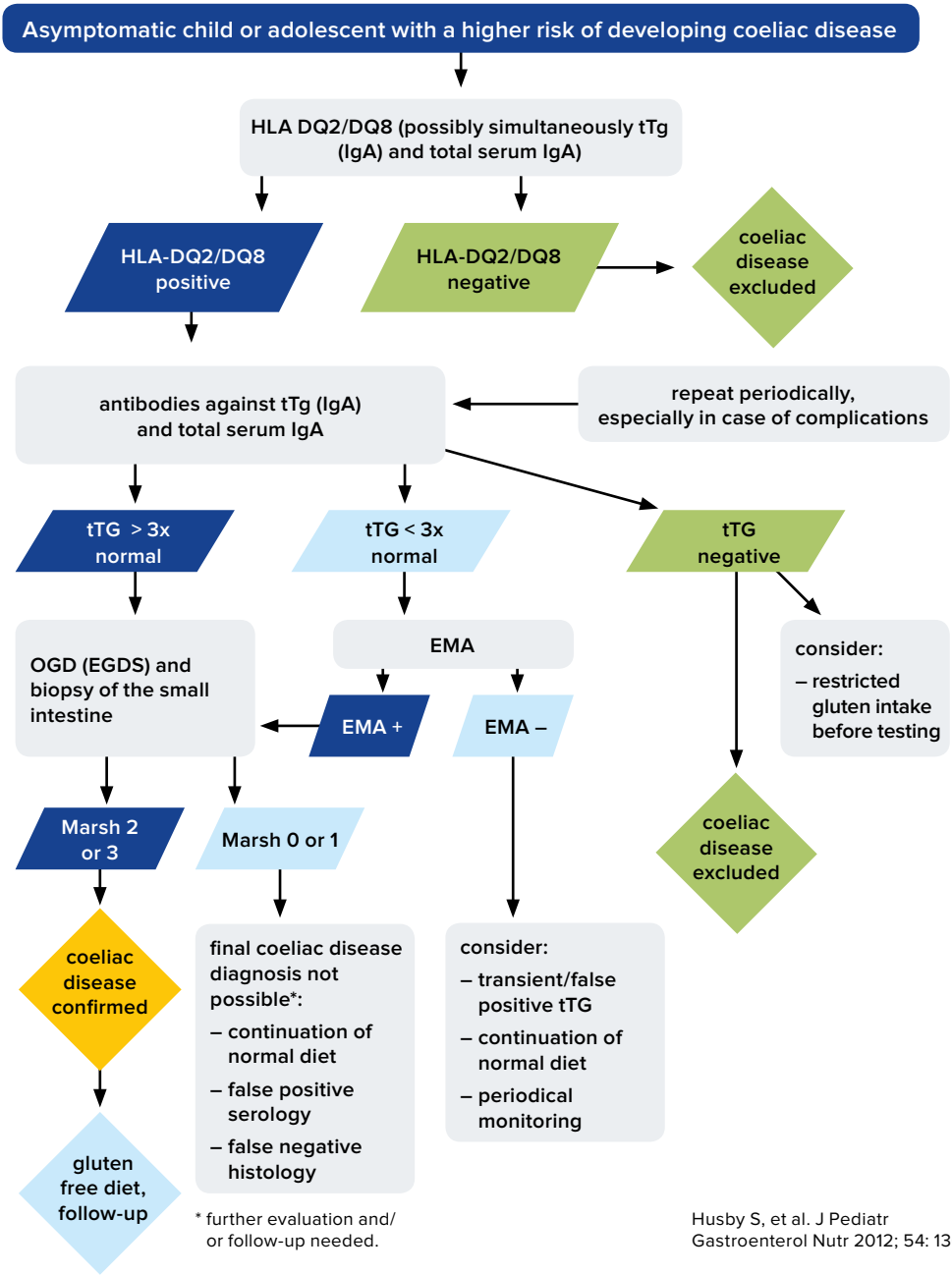
Igor, 47 years

# NO FEAR!

# Diagnostic approach in children and adolescents with symptoms or signs suggesting coeliac disease



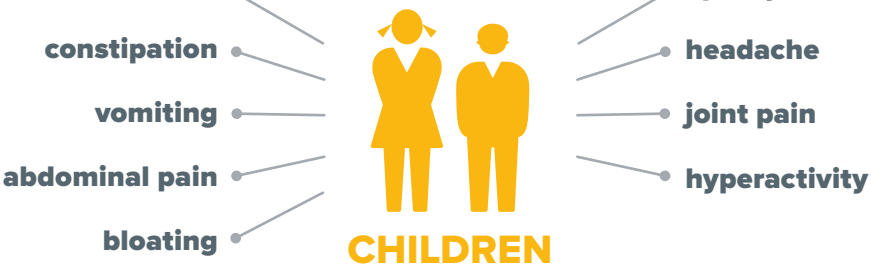
# Diagnostic approach in asymptomatic children or adolescents with a higher risk of developing coeliac disease




* further evaluation and/ or follow-up needed.

Husby S, et al. J Pediatr Gastroenterol Nutr 2012; 54: 136–60

# Symptoms





## Higher risk groups

first-degree relatives of coeliac disease patients
type 1 diabetes
immunoglobulin A deficiency
autoimmune thyroiditis
Down syndrome
Turner syndrome
Williams syndrome

# Signs

## CHILDREN

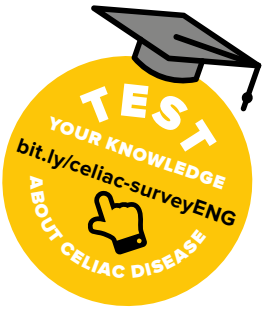
- malnutrition
- failure to thrive
- abdominal distension
- short stature
- anaemia
- dental enamel defects
- aphthae
- delayed puberty
- oedema
- rickets
- arthritis
- hypertransaminasaemia

## ADULTS

- brittle nails
- abdominal distension
- short stature
- iron deficiency anaemia
- oedema
- aphthae
- hair loss
- peripheral neuropathy
- malnutrition, weight loss
- myopathy
- hypertransaminasaemia
- osteopenia/osteoporosis
- bone fracture

# Complications

osteoporosis
autoimmune diseases
gynaecological disorders
haematological disorders
neurological disorders
psychiatric diseases
malignant lymphoma of the small intestine



# Glossary

***anaemia*** – a condition where the levels of haemoglobin (Hb) in blood are less than normal. One of the most frequent types of anaemia is the iron-deficiency anaemia (iron is required for Hb synthesis).

***antiendomysial antibodies (EMA)*** – antibodies (usually IgA class) directed against the endomysial tissue. These antibodies are usually present in the blood of coeliac disease patients in the active phase of the disease.

***antigliadin antibodies (AGA)*** – antibodies (IgA and IgG class) directed against gliadin. These antibodies may be present in the blood of coeliac disease patients in the active phase of the disease. In comparison to tTG and EMA their specificity is much lower.

***villous atrophy*** – a pathologic defect of the intestinal mucosa. The villi become shorter or even completely flattened (in the case of complete atrophy).

***biopsy*** – removal of a tissue sample in order to examine it in a variety of ways.

***gluten free diet*** – the only way to treat coeliac disease. A strict diet involves a complete avoidance of wheat, barley, rye and, in some coeliac disease patients, even oats.

***dermatitis herpetiformis*** – a skin condition characterised by itchy rash on the skin that appear on typical locations (e.g. elbows and knees). It is one of the possible clinical manifestations of coeliac disease.

***IgA*** – a subclass of antibodies (or immunoglobulins), found in blood and mucosal secretions. In case of IgA deficiency, coeliac disease is more common.

***intraepithelial lymphocyte*** – lymphocytes responsible for immunological protection, found between epithelial cells on the surface of the intestinal lining.

***lactose*** – milk sugar, composed of one glucose molecule and one galactose molecule. Lactose intolerance is common in coeliac disease and may disappear completely when a gluten-free diet is adopted.

***malabsorption*** – reduced absorption of nutrients as a consequence of digestive enzymes deficiency or damaged intestinal lining.

***antibodies*** – protein molecules capable of carrying out certain reactions, which usually have a protective function.

***antibodies directed against tissue transglutaminase (anti tTG)*** – antibodies (usually IgA class) directed against the tissue transglutaminase enzyme. These antibodies are usually present in the blood of coeliac disease patients in the active phase of the disease.

***villi*** – anatomical structures in the shape of "glove-fingers" typical for a normal intestinal lining.

***serological markers*** – antibodies, which can be detected in blood. Their presence represents a valuable diagnostic element in the detection of coeliac disease.

***HLA system*** – a complex of genes located on chromosome six, which are responsible for protein synthesis. Proteins play a crucial role in the immunological reaction.

***Higher-risk group*** – a group of people in the community with a higher-than-expected risk for developing a particular disease, which may be defined on a measurable parameter (e.g. an inherited genetic defect, physical attribute, lifestyle habit etc.)

***d-GP*** – deamidated gliadin peptide antibodies (dGP Ab) - antibodies (usually IgA class) directed against deamidated gliadin peptide. These antibodies are usually present in the blood of coeliac disease patients in the active phase of the disease.

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publisher: Mestna občina Maribor, authors: J. Dolinšek, J. Dolinšek, M. Klemenak, M. Karla, D. Sblattero, M. Milinović, T. Not, K. Werkstetter, T. Koltai, B. Koletzko, S. Koletzko, I. Korponay-Szabo, A. Mirkov, A. Likar Mastnak, T. Krenčnik, J. Gyimesi, S. Kostanjevec, design: Studio 8

This e-brochure will be regurarly updated based on new evidence based guidelines.