Coeliac Disease

Nursing Care and Management

Helen Griffiths MSc, RGN, Cert MHSC

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This book is dedicated to my father
whose belief
in me remains my beacon.
And my family who helps to keep it
blazing

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List of Contributors

Elaine Horne BSc, RGN
Clinical Nurse Specialist Gastroenterology,
Hereford Hospitals NHS Trust

James Horne,
Student

Patient Stories:
Alan Holmes
Zoe Verry
Jean Lloyd

Foreword

Helen has brought together years of experience of managing coeliac disease in this highly informative new book. It provides an excellent source of information for Specialist Nurses developing an up-to-date service for patients with coeliac disease and will also be a valuable source of information for patients and their families in exploring the many issues of this condition. The extensive literature search and detailed medical discussions will be of great interest to general practitioners and hospital specialists as the full range of holistic care of patients with coeliac disease is addressed. The multitude of real-life experiences that come out of every chapter make this a living account of the impact of coeliac disease on patients' every day lives and how all healthcare professionals can work together for the patients' benefit.

The individual chapters address the different aspects of coeliac disease in a clear but thorough manner with the final chapter of four true life stories bringing the most important issues together in patients' own words. Each of the preceding chapters have learning outcomes clearly laid out and also activities to improve the development of health services with particular emphasis on the type of patient information that is useful and practical.

The crucial question of accurate diagnosis is addressed not only from the scientific perspective of serological screening tests but also from how to undertake the initial and probably most important first consultation in the most productive way possible. The issue of the coeliac iceberg is developed with the 'below the waterline' concept introduced. Dietary management is considered as a medical nutrition

therapy with international standards of food labelling addressed in a thorough manner.

Practical advice of shopping lists for eating in is listed and good-humoured recommendations for eating out are given. The emotional impact of dealing with the diagnosis receives a rewarding amount of discussion and vulnerable groups in adolescence, pregnancy and old age are given specific consideration.

The team approach to the management of coeliac disease is considered essential with long-term aim of dietary compliance as the key to successful management. The option of telephone follow-up clinics is outlined but only if that is in the best interest of individual patients. The role of patient support groups and their benefits are discussed along with the issue of patient expectations.

Potential complications of osteoporosis, hyposplenism and risk of malignancy are carefully appraised with the evidence and current understanding of these complications described. Difficulties in management of resistant disease cover interesting possibilities of cross-contamination in addition to the rarer medical diagnoses of bacterial overgrowth, microscopic colitis and exocrine pancreatic insufficiency.

Future developments and current avenues of research are outlined with fascinating possibilities of dietary supplementation with endopeptidases, detoxification by transamidation of wheat flour and genetic modification of cereals to selectively reduce the toxic component of gluten.

This book has clearly been written to be enjoyed by a wide range of readers with the ultimate aim of improving the care and management of all patients with coeliac disease.

Dr Rupert A.J. Ransford, MRCGP, MD, FRCP
Consultant Gastroenterologist &
Clinical Director for Medicine
Hereford Hospitals
NHS Trust

Preface

It is thought that coeliac disease affects as many as 1:100 people in the United Kingdom, many of whom will remain undiagnosed.

The aim of this book is to provide nurses with the knowledge and understanding to not only help identify those possibly affected but understand the investigative pathway, the impact of the diagnosis and the long-term treatment and management.

The treatment of coeliac disease is the 'life-long' dietary exclusion of the protein gluten, found in wheat, barley, oats and rye. Anyone who has had their diet restricted voluntarily (weight watching) or involuntarily, even for a brief time, will know how attractive prohibited foods can be. Therefore, it should not be assumed that life-long dietary restriction is either easy or sustainable, but it is imperative if the long-term health risks associated with coeliac disease are to be avoided.

In my work as a nurse consultant and endoscopist I have come to appreciate the enormous impact that diagnosis and treatment can have on individuals with coeliac disease and the value of good support. This book is written to help nurses, through knowledge and understanding, and add value to the lives of those in their care with coeliac disease.

Introduction and How to Use This Book

'I read and I forget
I see and I remember
I do and I understand'
(Adapted from Confucius, 551–479 BC)

This quote will ring a salient bell with many of us; the busier we are, the less effectively we read and the less likely we are to remember what we have read unless it becomes a routine part of our lives. Hence, despite our best intentions when we sit down with a medical book meaning to enhance our knowledge on a subject, the chances are that we will skip bits or skim read and then still find ourselves a few weeks later saying 'I'm sure I read something about that'. Sound familiar?

The aim of this book is threefold:

- To increase nurses' knowledge and understanding of the care and management of coeliac disease.
- To enable general nurses to understand the health needs of coeliac patients in their care.
- To assist specialist nurses in developing services for coeliac patients.

To support these aims, each chapter has been designed to impart knowledge, but also to try and move you from simply reading to 'doing' something for coeliac patients and therefore understanding more fully the impact of this disease on individuals. In order to do this you will find a number of boxes within the text.

At the start of each chapter you will find a box with:

Intended learning outcomes, which will:
- Help you to make best use of each chapter
- Maximise your knowledge intake
- Help put theory into practice

The text is also interspersed with boxes highlighting:

Activities that will help you to:
Think about the information contained in each chapter and demonstrate how this might relate to and improve practice within your own clinical area.

In addition there are:

Hints and tips for service development:
With practical advice for nurses working within specialist areas who wish to provide dedicated supportive services for coeliac patients.

For specialist nurses hoping to develop a specific service for coeliac patients it is important to follow some basic principles which I hope you will find within this text:

- That the service is built on current evidence-based practice but remains responsive to growing evidence.
- That there are real benefits for patients including improved access to care at a time that is appropriate and timely and acknowledges coeliac disease as a long-term condition.
- That it is built around pathways and protocols that support the evidence base and audit its effectiveness.
- That links are maintained with all those involved in the service.
- That the personal and professional development and learning required in undertaking the role is acknowledged and built into the service.

Rather than detract from the readability of the text it is hoped that it will maximise your learning and lead to greater understanding.

Chapter 1

The History of Coeliac Disease

LEARNING OUTCOMES

At the end of this chapter you should be able to:

- Describe where the word 'coeliac' came from.
- Chart the early history of the disease.
- Critically discuss the current diagnostic criteria.

At the end of the last Ice Age, as we moved from the Mesolithic into the Neolithic era, people discovered that if they settled in one place for long enough, instead of relying on their hunter gatherer nomadic existence, they could sow and harvest crops of cereals like wheat.

One of the consequences of this Neolithic revolution was civilisation and the concept of production. Another, in all probability, was that people who could not tolerate wheat as part of their diet became ill.

We can surmise that coeliac disease has always existed. However, despite its first description as a clinical entity as far back as the second century AD, the fact that its numerous symptoms mimic several other conditions and that its cause has remained elusive has meant that its recognition as a distinct disorder, readily diagnosed and treated, has been a long and complicated journey. This is why history is so important, not only from the point of view of general interest but in helping both the healthcare profession and patients understand the complexity of the disease.

Activity

How do you think that the history of the disease might be useful when discussing the diagnosis with patients?
Consider here how you would feel if your diagnosis had been made after many years of feeling 'unwell'. Could the fact that our understanding has been slow be reassuring?

This is written with the understanding that when we talk of 'history' in coeliac disease, as more is known about the disease and its management even recent findings become history.

We start this chapter by describing the history of the disease, the history that led to the recognition of the environmental factors triggering the disease and the notable medical contributions made to its recognition as a distinct disorder. We then look at its definition and the challenges to the diagnostic criteria.

A CHRONOLOGICAL HISTORY OF COELIAC DISEASE

The second century AD

The word coeliac comes from the Greek word *koiliakos*, meaning 'suffering in the bowels'. This was the term used by Aretaeus of Cappadocia, a contemporary of the Roman Physician Galen in the second half of the second century AD. From his writings, edited and translated from Greek to Latin by Francis Adams and printed for the Sydenham Society of England in 1856 (Adams, 1856), koiliakos first became known as 'The Coeliac Affection'. *An affection of the digestion and of the distribution.*

In these translations we find the first descriptions of the disease, with features including fatty diarrhoea, pallor, weight loss and chronic relapse, affecting both children and adults. Aretaeus described the diarrhoea as being light in colour, offensive in odour and accompanied by flatulence. The patient

he described as emaciated and atrophied and incapable of performing any of his accustomed works.

The seventeenth century

Just a year prior to Adams' (1856) translation of Aretaeus' works, Dr Gull writing in Guy's hospital reports (Gull, 1855) outlined the case of a 13-year-old boy, whose symptoms of enlarged abdomen and frequent and voluminous stools of a dull, chalky colour clearly suggested the symptoms of coeliac disease as we understand them today. However, it was not until a few years later in 1888 that Dr Samuel Gee, using the same title as Francis Adams' (1856) translation, gave the second classic and first modern description of 'The Coeliac Affection' and laid the foundation not only for describing the condition, but also for establishing a criterion for its diagnosis and furthermore a recognition of its treatment being related to diet.

Gee (1888) described coeliac as a kind of chronic indigestion, affecting people of all ages but especially children between 1 and 5 years of age. As with Aretaeus, he describes a pale, loose, foul-smelling stool, frothy in appearance, resembling oatmeal porridge or gruel. He paints a vivid picture of a wasted almost cachectic patient, pale and puffy of face. He observes that unfortunately death is a common end and where patients survive, recovery is often incomplete, the illness dragging on for years with periods of relapse.

Dr Gee (1888) identified a causative link between the symptoms of emaciation, cachexia and diarrhoea and for the first time, rather prophetically, that (Gee, 1888):

> if the patient can be cured at all, it must be by means of diet (p. 20).

He made the observation that rice, sago and cornflour were unfit foods and that malted foods including rusks and bread were better. Although we now know the reverse to be true, for the first time Gee (1888) implied that unfit foods actually produced a pathologic condition of the digestive tract.

> What the patient takes beyond his power of digestion does harm (p. 20).

The twentieth century

The next important breakthrough in our understanding of coeliac disease did not come until 1908, as a culmination of 7 years of work between Dr Emmett Holt, senior director of children's medicine at Bellevue Hospital, and Christian Herter of Columbia University. They worked together on both the clinical and theoretical aspects of the disease and published their conclusions in a work entitled *On Infantilism from Chronic Intestinal Infection* (Herter, 1908). Their main observations were that this was a pathological state of childhood associated with a chronic intestinal infection. The chief manifestations of this intestinal infantilism were an arrest in the development of the body, without affecting mental development. It was, however, characterised by marked abdominal distension, a varying degree of anaemia, rapid onset of physical and mental fatigue, and irregularities of intestinal digestion resulting in frequent episodes of diarrhoea. Their most important contribution was the observation that whilst fats were tolerated moderately well, carbohydrates were poorly tolerated, almost always causing relapse or a return of diarrhoea. Whilst these conclusions were not universally accepted by colleagues, they did act as the catalyst for further research into the most effective dietary treatment.

Two assistants of Dr Holt, Dr John Howland and Dr Sidney Haas, took up the cause of their senior colleagues and in 1921, Dr Howland in his presidential address to the American Paediatric Society (Newland, 1921) presented a paper on *Prolonged Intolerance to Carbohydrates*. He observed that growth suffered in proportion to the length of time that symptoms persisted and that many children were as a consequence below the average in height. He again noted that of all the elements of food, carbohydrate was the one that had to be most rigorously excluded and that after initial improvement in symptoms, was the most difficult to add back into the diet. He particularly noted that bread and cereals were the last foods that could be