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EXECUTIVE SUMMARY

Introduction

Coeliac disease is an inappropriate immune response to the consumption of gluten, a protein that is found in wheat, rye, barley and oats.

It is recognized as an under-diagnosed life long disease estimated to affect up to 1% of the population, mostly people of Caucasian, Middle Eastern or West Asian descent (GESA, 2006). In Western Australia, the only reported data on prevalence is 0.4% (1:251) in a study of a rural population (Hovell & Collett, 2001).

Coeliac disease is characterized by chronic inflammation of the intestinal mucosa resulting in a variety of clinical manifestations that can substantially affect an individual’s health status and quality of life.

The consequences of untreated coeliac disease can be severe and may include an increased incidence of gastrointestinal cancer, infertility, osteoporosis, severe nutritional deficiencies and failure to thrive in children (Australian and New Zealand Coeliac Research Fund, 2007).

The only proven first-line effective treatment of coeliac disease is a life-long gluten-free diet.

Objectives of the Coeliac Disease Model of Care

The Coeliac Disease Model of Care is an articulation of best practice service provision across the continuum of care. It will provide a framework for the delivery of a person-centred approach that will ensure timely, effective, affordable and integrated service delivery for all Western Australians affected by this condition. The Coeliac Disease Model of Care is intended to facilitate the following:

- Increase health provider and consumer awareness of coeliac disease as an often poorly recognized and under diagnosed condition;
- Early diagnosis of coeliac disease by utilizing best practice guidelines and protocols;
- Provision of timely gastroscopy and distal duodenal biopsies to confirm histologic diagnosis when coeliac autoantibodies are positive;
- Coordinated and integrated management and care across clinical and support services;
- Preventing or managing associated complications of the condition that can substantially reduce quality of life; and
- Reduction in variations in screening, diagnosis and management that appear between different clinicians and allied health providers, across people from metropolitan, regional, rural and remote areas and in the care provided to disadvantaged groups.
The Coeliac Disease Model of Care covers the following stages:

- Community awareness of coeliac disease
- Early diagnosis of coeliac disease
- Best management and support following diagnosis of coeliac disease
- Best long term care and support

Summary of recommendations and outcomes

1. Increase awareness of coeliac disease
2. Ensure best practice guidelines for the screening, diagnosis and management of coeliac disease are available to all health professionals in the Western Australian health system.
3. Ensure the timely access (1-3 months) to endoscopy services to enable the early diagnosis of coeliac disease.
4. Ensure all people diagnosed with coeliac disease are provided with an action plan for effective ongoing management and self-management.
1. INTRODUCTION

Coeliac disease is an inappropriate immune response to the consumption of gluten, a protein that is found in wheat, rye, barley and oats\(^1\).

It is recognized as an under-diagnosed life-long disease estimated to affect up to 1% of the population, mostly people of Caucasian, Middle Eastern or West Asian decent (GESA, 2006). In Western Australia, the only reported data on coeliac disease prevalence is 0.4% (1:251) in a study of a rural population (Hovell & Collett, 2001).

Coeliac disease is characterized by chronic inflammation of the intestinal mucosa resulting in a variety of clinical manifestations that can substantially affect an individual’s health status and quality of life.

The consequences of untreated coeliac disease can be severe and may include an increased incidence of gastrointestinal cancer, infertility, osteoporosis, severe nutritional deficiencies and failure to thrive in children (Australian and New Zealand Coeliac Research Fund, 2007).

The only proven first-line effective treatment of coeliac disease is a life-long gluten-free diet.

The awareness of ‘gluten free’ as a dietary regime and commercially available gluten free products is increasing. Increased community awareness is needed to ensure that diagnosis is made and a rigid dietary regime is promoted to avoid long-term complications from poor control.

\(^1\) The inclusion of oats in a gluten free diet is controversial and is generally not recommended. For further specific advice, consultation with an accredited practicing dietitian or gastroenterologist is recommended.
2. MODEL OF CARE OVERVIEW

The Coeliac Disease Model of Care is an articulation of best practice for those diagnosed with coeliac disease across the continuum of patient care. It covers both adults and children.

Coeliac Disease has been selected as a priority digestive health condition because it is an often poorly recognised and under diagnosed condition that affects the health and quality of life of a significant number of people.

2.1 Model of Care Stages and Key Objectives

<table>
<thead>
<tr>
<th>Stages across the coeliac disease continuum</th>
<th>Key Objectives</th>
</tr>
</thead>
<tbody>
<tr>
<td>Community awareness of coeliac disease</td>
<td>Ensure coeliac disease awareness strategies are in place.</td>
</tr>
<tr>
<td>Diagnose coeliac disease early</td>
<td>Dissemination of information and the application of best practice diagnostic protocols</td>
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<td>Best management and support following diagnosis of coeliac disease</td>
<td>Development of a care plan for all people diagnosed with coeliac disease</td>
</tr>
<tr>
<td>Best long term care and support</td>
<td>Optimal management and self management of coeliac disease to maintain good health and quality of life.</td>
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</table>

2.2 Methodology

A partnership between the Clinical Lead of the Digestive Health Network Executive Advisory Group and The Coeliac Society of Western Australia (CSWA) was established in March 2007 to commence formative work on the development of a Coeliac Disease Model of Care.

This partnership commenced reviewing data, literature and existing models of coeliac disease diagnosis and management. This formative work was utilized to commence the development of the Coeliac Disease Model of Care document by the Health Networks Branch with ongoing input from the Digestive Clinical Lead and the Executive Officer of the CSWA.

Further expert consultation through members of the Digestive Health Network Executive Advisory Group occurred throughout the development of the model of care. Feedback and submissions to iterations of the model were considered and incorporated as appropriate. Specific input was also sought from representatives from the Australia and New Zealand Coeliac Research Fund, paediatric and allied health professionals and from the Endocrine and Infections & Immunology Health Networks. The final draft was then circulated to the wider digestive health network for comment.
The Digestive Health Network Executive Advisory Group endorsed the finalised model of care. The executive advisory group comprises:

<table>
<thead>
<tr>
<th>Name</th>
<th>Role</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dr Andre Chong</td>
<td>Clinical Lead and Chair</td>
</tr>
<tr>
<td>Professor David Fletcher</td>
<td>Surgery and Pathology, Fremantle Hospital and UWA</td>
</tr>
<tr>
<td>Ms Iren Hunyadi</td>
<td>Consumer representative</td>
</tr>
<tr>
<td>Professor John Olynyk</td>
<td>Academic and research representative UWA</td>
</tr>
<tr>
<td>Dr John Lindsey</td>
<td>WA Country Health Services representative</td>
</tr>
<tr>
<td>Dr Ian Lawrance</td>
<td>Gastroenterologist, Fremantle Hospital</td>
</tr>
<tr>
<td>Dr Hooi Ee</td>
<td>Gastroenterologist, Sir Charles Gairdner Hospital</td>
</tr>
<tr>
<td>Dr Wendy Cheng</td>
<td>Gastroenterologist and hepatologist, Royal Perth Hospital</td>
</tr>
<tr>
<td>Ms Amanda McKnight</td>
<td>Clinical Nurse Specialist, Sir Charles Gairdner Hospital</td>
</tr>
<tr>
<td>Ms Suzanne Tillotson</td>
<td>Perth &amp; Hills Division of General Practice</td>
</tr>
<tr>
<td>Ms Samantha Boggs</td>
<td>Dietitians Association of Australia and Joondalup Health Campus</td>
</tr>
<tr>
<td>Dr Susan Stevenson</td>
<td>General Practitioner</td>
</tr>
</tbody>
</table>
3. CURRENT STATE OF PLAY FOR COELIAC DISEASE

3.1 Definition of Coeliac Disease

Coeliac disease is an inappropriate immune response to the consumption of gluten, a protein that is found in wheat, rye, barley and oats (Australian and New Zealand Coeliac Research Fund, 2007).

In the presence of dietary gluten, coeliac disease is characterised by a chronic inflammatory state of the proximal small intestinal mucosa, which can impair digestion resulting in malabsorption of nutrients and cause a variety of clinical manifestations.

Table 1. Clinical manifestations of coeliac disease

<table>
<thead>
<tr>
<th>Fatigue</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iron deficiency and anaemia</td>
</tr>
<tr>
<td>Altered bowel habits (including diarrhoea and/or constipation)</td>
</tr>
<tr>
<td>Abdominal pain or bloating</td>
</tr>
<tr>
<td>Flatulence</td>
</tr>
<tr>
<td>Irritability and depression</td>
</tr>
<tr>
<td>Nutritional deficiency</td>
</tr>
<tr>
<td>Weight loss or gain</td>
</tr>
<tr>
<td>Pubertal delay, psycho-motor delay, growth failure and failure to thrive (in children)</td>
</tr>
<tr>
<td>Bone or joint pain and stiffness</td>
</tr>
<tr>
<td>Dental enamel defects</td>
</tr>
<tr>
<td>Mouth ulcers</td>
</tr>
<tr>
<td>Itchy and blisterly skin rash</td>
</tr>
<tr>
<td>Undefined neurological disorder/epilepsy</td>
</tr>
</tbody>
</table>

(NIH, 2004)

Untreated coeliac disease may lead to an increased risk of gastrointestinal malignancy, vitamin and mineral deficiencies, osteoporosis and other extra intestinal problems (GESA, 2006). In children, consequences include poor growth and development and delayed puberty (North American Society for Pediatric Gastroenterology, Hepatology and Nutrition, 2005).

Coeliac disease is not an allergy and should not be confused with wheat intolerance or wheat allergy (Australian and New Zealand Coeliac Research Fund, 2007).

In a small subgroup of patients, clinical and histological abnormalities persist or recur despite a strict gluten free diet. This non-responsiveness is known as refractory coeliac disease and is due to an unexplained sustained stimulation of T cell activity. This may develop into enteropathy associated T-cell lymphoma and requires further investigation by a specialist/gastroenterologist (AGA, 2006b).
3.2 Prevalence of Coeliac Disease

Coeliac disease is recognized as an under-diagnosed life long disease estimated to affect 0.3-1% of the Australian population (63,000-210,000). (GESA, 2006) In Western Australia coeliac disease prevalence data is limited. The only reported data is 0.4% (1:251) in a study of a rural population (Hovell & Collett, 2001). However, only around 1 in 5 people with coeliac disease are thought to have been diagnosed (ANZCRF, 2007).

3.3 Morbidity & Mortality

Untreated patients may have an increased risk of malignancies. These include small bowel adenocarcinoma, enteropathy associated T-cell lymphoma and oesophageal and oropharyngeal squamous cell carcinoma (AGA, 2006a).

Morbidity from undiagnosed coeliac disease is due to ongoing symptoms including lethargy, altered bowel habits, neuropsychiatric disorders, infertility, failure to thrive, weight loss, steatorrhoea, iron deficiency, anaemia, nutritional deficiencies, metabolic bone disease, lymphoma and gastrointestinal disorders.

3.4 Risk Factors for Coeliac Disease

The risk of coeliac disease is increased for individuals identified in Table 2.

Table 2. Conditions associated with an increase prevalence of coeliac disease (prevalence rates are in brackets)

- 1st degree relative coeliac disease (10%)
- 2nd degree relative coeliac disease (2.6 - 5%)
- T1 Diabetes Mellitus (2 - 5% in adults, 3-8% in children)
- Down’s syndrome (3 - 12%)
- Turner’s syndrome (2 - 10%)
- Autoimmune thyroid disease (1.5 - 6.7%)
- Liver disease (Abnormal transaminases 1.5-9%, autoimmune hepatitis 2.9-6.4% and cirrhosis up to 6%)
- Osteoporosis (1 - 3%)
- Unexplained Infertility (2.1 - 4.1%)

(AGA, 2006a)

Testing for coeliac disease should be considered, especially where symptoms are present, in conditions identified above and Sjogrens Syndrome, unexplained recurrent fetal loss, unexplained delayed puberty, selective IgA deficiency, irritable bowel syndrome, peripheral neuropathy, cerebella ataxia and recurrent migraine, as well as children with short stature and first and second degree relatives of patients with coeliac disease.

3.5 Diagnosis of Coeliac Disease

The recommended screening test for coeliac disease is an assay of serum IgA autoantibodies to tissue transglutaminase g (IgA tTG antibodies)² (AGA, 2006b). It is recommended that serum IgA concentrations are requested at
the same time, as selective IgA deficiency is more common in patients with coeliac disease and, if present, will lead to false negative results (Anderson, 2005). The prevalence of coeliac disease in patients with IgA deficiency is recognized up to 8%, while 1.7-3% of patients with coeliac disease may be IgA deficient (AGA, 2006a). In patients with IgA deficiency, IgG tTG autoantibodies or IgG EMA (Endomysial antibodies) can be used in diagnosis (Green and Jabri, 2006).

The ‘gold standard’ for the diagnosis of coeliac disease is histology from the distal duodenum (AGA, 2006a). In both children and adults with positive coeliac autoantibodies, referral to a gastroenterologist (to obtain distal duodenal biopsies with gastroscopy and for further management) is recommended. As histologic changes in coeliac disease can be patchy to diagnose or confidently exclude coeliac disease, a minimum of 4-6 distal duodenal biopsies is recommended (AGA, 2006a).

2 In an adult population, the pooled estimates of the sensitivity and specificity of IgA tTGA-HU were 95.1% (95% CI, 91.8%-98.1%) and 98.3% (95% CI, 97.1%-99.6%), respectively. Among the studies in children, the pooled estimates of sensitivity and specificity were 95.7% (95% CI, 94.6%-99.8%), respectively. (AGA 2006a)
**Histologic grading in coeliac disease**

Histologic changes can be graded according to severity from 0 to 4, based on Marsh’s grading system (Table 3).

**Table 3. Marsh’s Grading System**

<table>
<thead>
<tr>
<th>Marsh stage 0: Normal mucosal and villous architecture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marsh stage 1: Infiltrative</td>
</tr>
<tr>
<td>Normal mucosal and villous architecture</td>
</tr>
<tr>
<td>Increased number of intra-epithelial lymphocytes</td>
</tr>
<tr>
<td>Marsh stage 2: Hyperplastic</td>
</tr>
<tr>
<td>Similar to above, but with enlarged crypts and with</td>
</tr>
<tr>
<td>Increased crypt cell division</td>
</tr>
<tr>
<td>Marsh stage 3: a. Partial villous atrophy</td>
</tr>
<tr>
<td>Shortened blunt villi</td>
</tr>
<tr>
<td>Mild lymphocyte infiltration</td>
</tr>
<tr>
<td>Enlarged hyperplastic crypts</td>
</tr>
<tr>
<td>b. Subtotal villous atrophy</td>
</tr>
<tr>
<td>Clearly atrophic villi, but still recognizable</td>
</tr>
<tr>
<td>Enlarged crypts whose immature epithelial cells are</td>
</tr>
<tr>
<td>generated at an increased rate</td>
</tr>
<tr>
<td>Influx of inflammatory cells</td>
</tr>
<tr>
<td>c. Total villous atrophy</td>
</tr>
<tr>
<td>Complete loss of villi</td>
</tr>
<tr>
<td>Severe crypt hyperplastic, and infiltrative inflammatory lesion</td>
</tr>
<tr>
<td>Marsh stage 4: Hypoplastic</td>
</tr>
<tr>
<td>Total villous atrophy</td>
</tr>
<tr>
<td>Normal crypt depth, but hypoplasia</td>
</tr>
<tr>
<td>Normal intraepithelial lymphocyte count</td>
</tr>
<tr>
<td>Many feel this does not exist and represents severe</td>
</tr>
<tr>
<td>malnutrition</td>
</tr>
</tbody>
</table>

(AGA, 2006a)

Marsh Stage 3 and above is consistent with characteristic histopathologic features of coeliac disease. As histologic changes are not specific for coeliac disease diagnosis in patients with Marsh Stage 1 or 2 changes needs further supportive evidence including clinical manifestations, serology or HLA testing (AGA, 2006a). In coeliac disease the histologic changes classically improve or reverse after gluten is removed from the diet.

Patients need to be on an adequate gluten containing diet prior to both screening, testing, and biopsy. In both children and adults, the recommended amount of gluten ingestion prior to testing is 10g of gluten/day (4-5 slices of white bread). In adults this should be over a duration of at least 4 weeks (BSG, 1998). A gluten free diet should not be commenced until diagnosis is confirmed.

Screening of some individuals in very high risk groups (Table 2) is

**In children belonging to high-risk groups, testing is recommended from the age of 3.** These children need to be on an adequate gluten containing diet for at least one year (10g of gluten/day ie 4-5 slices of white bread). Prior to the age of 3, coeliac autoantibody tests are unreliable (NASPGH, 2005). Screening should also be undertaken if symptoms compatible with coeliac disease develop.
recommended on an annual basis as there is evidence that individuals may develop a positive test on repeat testing, and have biopsies compatible with coeliac disease even if they have previously tested negative.

Genotype testing for HLA-DQ2 and HLA-DQ8 may be useful in some cases, including asymptomatic children in high risk groups (NASPGHAN, 2005). Almost all patients with coeliac disease have HLA-DQ2 (~ 95% of patients with coeliac disease) or HLA-DQ8 (~ 5% of patients with coeliac disease), and so absence of these excludes coeliac disease and thus can be useful in excluding the disease when autoantibody tests are indeterminate. Presence of the HLA-DQ2 and HLA-DQ8 genotypes have a poor specificity for predicting coeliac disease as they are common within the general population (AGA, 2006a).

If the clinical suspicion is high, despite negative serology, in both children and adults, referral should be made to a gastroenterologist for consideration of HLA-DQ typing and/or a gastroscopy with distal small bowel biopsies (AGA, 2006a).

Some patients may be ‘gluten sensitive/intolerant’ and exhibit symptoms such as bloating and altered bowel habit, but have negative serology and normal duodenal biopsies. These patients do not have coeliac disease but may choose to be on a gluten free diet for symptomatic control.

The screening and diagnostic pathway for coeliac disease is described in Appendix 1.

3.6 Management of Coeliac Disease

Commencing and maintaining a strict, lifelong gluten free diet is the cornerstone of managing coeliac disease (AGA, 2006a).

Patients should be seen at least once by an experienced clinician, such as a gastroenterologist, for accurate information and an action plan, once a confirmatory histological diagnosis is available. Nutritional deficiency states should be looked for and corrected accordingly. Assessment for osteoporosis by bone mineral density measurement should be performed in adults regardless of age or gender. Patients should also be referred to an Accredited Practicing Dietitian and support organisation, such as The Coeliac Society of WA, for additional information and support. Patients should be followed up by their primary physicians for compliance and early detection of refractory coeliac disease or complications from the disease.

Seventy percent of symptomatic patients have a noticeable clinical improvement within 2 weeks. Patients who remain symptomatic (e.g. persistent weight loss, malabsorption, iron deficiency anaemia, abdominal pain or night sweats) after a few weeks on a strict gluten free diet may have refractory coeliac disease and should be referred to a gastroenterologist for further management.

Repeat testing of coeliac antibodies (IgA tTG) after 6 months of a gluten free diet should demonstrate a decrease of antibody titre. Annual serologic testing can identify a rise or persistent elevation of titre that requires further review. This should include referral to a gastroenterologist to
exclude refractory coeliac disease and a dietitian to assess dietary compliance and education.

Some parents of young children may need additional psychological support in adjusting and managing challenging early childhood behaviour that threatens compliance with a gluten free diet.

The management pathway for coeliac disease is described in Appendix 2.
4. CURRENT SERVICE PROVISION

Current health service provider awareness, diagnostic practices and management protocols for coeliac disease are variable in Western Australia. This is linked to gaps in provider education across primary care, access to endoscopy, specialist and dietitian services and the need for agreed approaches in the development of action plans and self-management protocols. These issues are of particular significance in the provision of coeliac disease diagnosis and management services in rural and remote areas of Western Australia.

4.1 Primary Care Services

Primary care services currently play the principal role in effecting the diagnosis and management of coeliac disease.

- Identification of possible coeliac disease by GPs, dietitians, community and child health professionals
- Screening for coeliac disease by GPs
- Referral to specialist services, public and private.

4.2 Public Hospital Services

The following coeliac disease services are provided through the public hospital system.

- Triaging of referrals
- Provision of endoscopy and pathology services
- Screening for coeliac disease by medical staff for identified in/outpatients.
- Gastroenterologist and dietitian consultation

a) Tertiary Public Hospitals

Fremantle Hospital, Sir Charles Gairdner Hospital, Royal Perth Hospital and Princess Margaret Hospital manage coeliac disease patients through general gastroenterology clinics. This includes the provision of dietetic services.

b) General Public Hospitals

No specific clinics are available in general public hospitals

Inpatient Services

The identification and appropriate care of inpatients with coeliac disease is variable in the public hospital system. This particularly applies to the provision of gluten-free food and suitable ingested medications.

Outpatient Services

- Screening, triaging of referral and ongoing management of patients with coeliac disease.
- Dietetic services - public, private, relevant NGO services.
4.3 **Gastroenterology clinics in private hospitals**

Some private hospitals provide the following services for coeliac disease patients through dedicated gastroenterology clinics.

- Triaging of referrals
- Provision of endoscopy and pathology services
- Screening for coeliac disease by medical staff for appropriate in/outpatients.
- Gastroenterologist and dietitian consultation

4.4 **Non-government organisations**

Provision of education and information on coeliac disease and a gluten free diet for people with coeliac disease, their families and carers is provided by non-government organizations such as The Coeliac Disease Society of WA. This also includes some disease management and self-management support and the provision of private dietitian services.

4.5 **Country (rural and remote) services**

There is limited access to screening, diagnostic and management services for coeliac disease in rural and remote areas. Areas of deficiency include availability and access to the following services:

- GP services
- Endoscopy services
- Specialist (gastroenterology) services
- Dietetic services
- Education and disease management support
5. FUTURE MODEL OF CARE FOR COELIAC DISEASE

The Coeliac Disease Model of Care is an articulation of best practice service provision across the continuum of care. It will provide a framework for the delivery of a person-centred approach that will ensure timely, effective, affordable and integrated service delivery for all Western Australians affected by this condition. The model of care covers four key stages.

1. Community Awareness of Coeliac Disease
2. Early diagnosis of coeliac disease
3. Best management and support following diagnosis
4. Best long term care and support

5.1 Stage 1: Community Awareness of Coeliac Disease

Key Objective: Coeliac disease awareness strategies are in place.

Recommendations

- That WA Area Health Services support community initiatives on the awareness of coeliac disease.
- That appropriate non-government organisations, in collaboration with WA Health and local gastroenterologists with a specific interest in coeliac disease, be contracted to develop coeliac disease awareness and information strategies.

5.2 Stage 2: Early Diagnosis of Coeliac Disease

Key Objective: Dissemination of information and application of best practice diagnostic protocols

Recommendations

- WA Health will ensure best practice information and protocols for the screening and diagnosis of coeliac disease be made available to all health professionals across WA.
- That screening for coeliac disease in appropriate patients be carried out by GPs, Paediatricians, Gastroenterologists or Internal Medicine Physicians (public and private) using best practice diagnostic protocols and guidelines.
- See Appendix 1
- That pathology services across the state will provide information to GP’s to reinforce that positive screening tests indicate coeliac disease is likely and diagnosis should be confirmed by small bowel biopsy. This will also include information emphasising that a gluten free diet should not be started before a small bowel biopsy is taken.
- Provide individuals with positive screening tests for coeliac disease access to gastroscopy (and distal duodenal biopsies) within one to three months for histological diagnosis. Gastroscopies will be performed by accredited Gastroenterologists, Surgeon Endoscopists or Internal Medicine Physicians.
- See Appendix 1
All health professionals will reinforce best practice information and protocols for the screening, diagnosis and management of coeliac disease.

5.3 Stage 3: Best Management and Support Following Diagnosis

Key Objective: Development of a care plan for all people diagnosed with coeliac disease

Recommendations

- Best practice information and protocols for the management of coeliac disease will be available to all health professionals on a DOH website.
- Gastroenterologists will provide a care plan to individuals diagnosed with coeliac disease. This will involve shared care with GPs. This includes identification of any nutritional deficiencies or associated conditions and referral to an Accredited Practising Dietitian or an appropriate non-government organisations offering coeliac disease management support. GP’s will be encouraged to consider the use of the EPC (Enhanced Primary Care program) to enable individuals to access private dietetic services. Patients will be encouraged to have their own dossier of investigations results such as serology, histology, biochemistry and bone densitometry (DEXA) results.
- Individuals with newly diagnosed coeliac disease will be tested for nutritional deficiencies and other associated conditions or complications. This includes:
  - Bone density;
  - Thyroid function;
  - Undiagnosed anaemia;
  - Nutrient deficiency (eg B12, Folate, Vit D); and
  - Psychological issues.
- WA Health, in partnership with appropriate non-government organisations and other relevant groups will develop, promote and support patient Self-management services.
- Health services will develop mechanisms for distribution of the patient information package to all newly diagnosed patients in collaboration with appropriate non-government organisations and other relevant community groups.
- Patients with nutritional deficiencies and other associated conditions or complications will have individual management plans.
- The following providers will adopt a self-management approach:
  - GPs;
  - Gastroenterologists, other relevant Internal Medicine Physicians and Paediatricians;
  - Hospital outpatients;
  - Dietitians;
  - Allied health services;
  - Community and child health services; and
  - Appropriate non-government organisations.
5.4 Stage 4: Best Long-Term Care and Support

Key Objective: Optimal management and self-management of coeliac disease to maintain good health and quality of life.

Recommendations

- Provide best practice information and protocols for the long-term management of coeliac disease
- Recall systems that support periodic assessment for individuals diagnosed with coeliac disease will be put in place by GP’s and/or gastroenterologists
- People who develop complications of coeliac disease will receive timely, appropriate management
- Individuals diagnosed with coeliac disease will have access to dietetic services through:
  - Department of Health Services, including tertiary, secondary and community settings
  - Private Accredited Practising Dietitians (through the use of Enhanced Primary Care plans by GPs)
- Referral by the GP to a gastroenterologist if refractory coeliac disease is suspected.
- Establish or improve links between country health care providers and metro-based expertise including the employment of telemedicine and other technologies.
- Appropriate non-government organisations and other relevant stakeholders (including WA gastroenterologists with a special interest in coeliac disease), will work collaboratively to:
  - Implement a statewide health professional training program for the management of coeliac disease (including metropolitan, regional, rural and remote localities).
  - Establish formal linkages to provide disease management support to country providers.
- Form local coeliac disease management support groups in the metropolitan area and larger rural centres/towns:
  - Through non-government organisations; and
  - Through community dietitians/allied health services.
- All people in WA with coeliac disease will have access to high quality services from health care providers and access to other trained health professionals, including dietitians (with a specific interest in coeliac disease), as needed.
- Facilitate the development of at least one or more specialised coeliac disease clinics in one or more tertiary teaching hospitals in WA.
6. OTHER CONSIDERATIONS

6.1 Workforce
Increasing the skills and capacity of the health workforce in coeliac disease diagnosis and management is an area that needs to be addressed. This will include:

- Appropriate Area Health Service staff requiring upskilling on coeliac disease will be identified as part of the performance development process and released for in-service training opportunities; and

- Health service resources are reviewed to support the workforce allocation for the management of coeliac disease, particularly specialised gastroenterology and dietetic services.

6.2 Horizon Scan
Research has commenced on the development of a non-dietary therapeutic ‘vaccine’ for the management of coeliac disease. Phase I clinical trials are due to begin in 2008.

7. SUMMARY OF RECOMMENDATIONS

- Increase awareness of coeliac disease
- Ensure best practice guidelines for the screening, diagnosis and management of coeliac disease are available to all health professionals in the Western Australian health system.
- Ensure the timely access (within one to three months) to endoscopy services to enable the early diagnosis of coeliac disease.
- Ensure all people diagnosed with coeliac disease are provided with an action plan for effective ongoing management and self-management.

8. EVALUATION

WA Area Health Services and Health Networks will work together to develop Key Performance Indicators to evaluate the implementation of the Coeliac Disease Model of Care.
REFERENCES


BIBLIOGRAPHY

Australian and New Zealand Coeliac Research Fund (2007). Coeliac Disease For the Public - information paper/CD


Department of Health (2005). Delivering a Healthy WA Strategic Intent 2005 - 2010, Perth, Western Australia, Department of Health WA.


The Coeliac Society of Western Australia (2007). Diagnosis of Coeliac Disease - brief consumer information paper.

APPENDICES

Appendix 1: Diagnosis of Coeliac Disease

Suspicion of coeliac disease: see symptoms and associated conditions

Request IgA tTG (tissue transglutaminase) & IgA (NB serology unreliable if already on gluten free diet)

If IgA deficient, IgG tTG/IgG EMA

If (+) serology, or persisting symptoms refer to gastroenterologist for histologic diagnosis (gold standard).

*Do not start on gluten free diet prior to histologic diagnosis as this may make confirmation of disease difficult*

In children <3 years serology may not be reliable and referral for histological diagnosis should be made as clinically indicated.

Triage to enable histological diagnosis to occur within 1-3 months of (+) serology or persisting symptoms

Please refer to Appendix 2 for management plan

Gastroenterology 2006;131:1977-1980

Common symptoms

- Fatigue
- Iron deficiency and anaemia
- Altered bowel habits (including diarrhoea and/or constipation)
- Abdominal pain or bloating
- Flatulence
- Irritability and depression
- Nutritional deficiency
- Weight loss or gain
- Pubertal delay, psycho-motor delay, growth failure and failure to thrive (in children)
- Bone or joint pain and stiffness
- Dental enamel defects
- Mouth ulcers
- Itchy and blistered skin rash

Conditions associated with increased prevalence of coeliac disease

- 1st degree relative coeliac disease
- 2nd degree relative coeliac disease
- T1 Diabetes Mellitus
- Down’s syndrome
- Turner’s syndrome
- Autoimmune thyroid disease
- Liver disease
- Osteoporosis
- Infertility/recurrent miscarriage
- Peripheral neuropathy, cerebella ataxia and recurrent migraine
- Short stature in children
### Appendix 2: Management of Coeliac Disease

**Confirmed histology and serology**

**Referral to**
1. Dietitian - nutritional assessment, gluten free diet education
2. Coeliac Society - support and development of self management skills
3. Gastroenterologist - for management plan and identification/management of any complications

**Recommended serologic screening in 1st degree (10% prevalence) and symptomatic 2nd degree (2.5-5%) relatives**

**Bone densitometry (DEXA scan)**
- FBC, LFTs, Fe studies, B12, Folate, TSH, Ca, PO₄, Vit D
- Correct any deficiencies (consider referral to endocrinologist if required)

**Repeat IgA tTG in 1 yr (may normalise after gluten free diet for >6mths)**
- Repeat histology in 1 yr (if still symptomatic or persistently elevated IgA tTG)

**Ongoing follow up by GP - encourage compliance with gluten free diet**

**Refer to gastroenterologist if recurrent/unresolving symptoms**
- (refractory coeliac, lymphoma, lactose/fructose intolerance, other diagnoses)
Appendix 3: Matrices and Summary Tables for Each Stage of the Model of Care

Table parameters:

**What** should be provided?
- What are the specific actions that should be undertaken?
- What activities, programs and services should be delivered?
- What are the guidelines and protocols that should be in place?
  - What needs to be done to improve integration within and between the stages?
  - What policy should be in place to support this?

**Who** should provide it?
- Who should comprise the care providers?
- Who should lead the care providers?
- What competencies are required to deliver the care?
- What training and education requirements are there?
- What policy should be in place to support this?

**Where** should it be provided?
- Where should the various activities/services/programs be provided?
  - eg in community, primary care, outpatient care settings?
- What policy should be in place to support this?

**Policy**
- Policy sources for each stage.
### STAGE 1. COMMUNITY AWARENESS OF COELIAC DISEASE

<table>
<thead>
<tr>
<th>WHAT</th>
<th>WHO</th>
<th>WHERE</th>
<th>POLICY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Awareness of signs and symptoms</td>
<td>DOH</td>
<td>General Practice</td>
<td>General Practitioners</td>
</tr>
<tr>
<td>Awareness of groups at increased risk</td>
<td>Area Health Services and hospitals</td>
<td>Community and child health centres</td>
<td>Aust &amp; NZ Coeliac Research fund</td>
</tr>
<tr>
<td>Importance of early diagnosis</td>
<td>Community and child health centres</td>
<td>Area Health Services and hospitals</td>
<td>NGO</td>
</tr>
<tr>
<td>Equitable availability of information</td>
<td>General Practice</td>
<td>NGO</td>
<td>Professional associations - ADA, AGA, GESA</td>
</tr>
<tr>
<td>Public awareness strategies</td>
<td>Allied health services - dietetics</td>
<td>Naturopathy clinics</td>
<td>DOH/Health Networks</td>
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<tr>
<td></td>
<td>Naturopathy</td>
<td>Community pharmacies</td>
<td></td>
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<tr>
<td></td>
<td>NGO</td>
<td>Professional associations</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Other consumer organisations</td>
<td>Community meeting halls</td>
<td></td>
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<tr>
<td></td>
<td>Community pharmacies</td>
<td>Media</td>
<td></td>
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<td></td>
<td>Media organisations</td>
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</table>
STAGE 2. DIAGNOSE COELIAC DISEASE EARLY

<table>
<thead>
<tr>
<th>WHAT</th>
<th>WHO</th>
<th>WHERE</th>
<th>POLICY</th>
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<tbody>
<tr>
<td>Screening and diagnostic tests will be performed according to best practice evidence.</td>
<td>Screening by: GPs, gastroenterologists, surgeon endoscopists and internal medicine physicians - public and private. (See Appendix 1)</td>
<td>Area Health Services and hospitals</td>
<td>General Practitioners</td>
</tr>
<tr>
<td>People throughout WA will have timely access to screening and diagnostic services, including:</td>
<td>Diagnosis by gastroenterologists, surgeon endoscopists and internal medicine physicians - public and private. (See Appendix 1)</td>
<td>Community and child health centres</td>
<td>Gastroenterologists/ Specialists - public and private</td>
</tr>
<tr>
<td>- Pathology</td>
<td></td>
<td>General Practice and GP Divisions</td>
<td>Hospitals - public and private</td>
</tr>
<tr>
<td>- Endoscopy</td>
<td></td>
<td>Gastroenterology/</td>
<td>Pathology services</td>
</tr>
<tr>
<td>- Gastroenterologist advice and/or consultation</td>
<td></td>
<td>specialist services - public and private</td>
<td>NGO - Coeliac Society of WA DOH</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pathology services - public and private</td>
<td>Profesional associations - ADA, AGA, GESA</td>
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</table>
## STAGE 3. BEST MANAGEMENT AND SUPPORT FOLLOWING DIAGNOSIS OF COELIAC DISEASE

<table>
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<tr>
<th>WHAT</th>
<th>WHO</th>
<th>WHERE</th>
<th>POLICY</th>
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</thead>
<tbody>
<tr>
<td>All people in WA with newly diagnosed coeliac disease and their families will have access to information about their condition and process for optimal management.</td>
<td>General Practice</td>
<td>General Practice</td>
<td>General Practice</td>
</tr>
<tr>
<td>Consistent, best practice care will be provided to individuals diagnosed with coeliac disease.</td>
<td>GPs, Gastroenterologists, Dietitians, Physicians, Paediatricians</td>
<td>Gastroenterology/specialist clinics - public and private, Hospital outpatients, Community &amp; child health centres, Allied health/dietetic services, NGO</td>
<td>Gastroenterology, Allied health/dietetics, NGO, DOH/Health Networks, Professional associations - ADA, AGA, GESA</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Organisation</th>
<th>Individual</th>
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<tbody>
<tr>
<td>General Practice</td>
<td>GPs</td>
<td></td>
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<tr>
<td>Gastroenterology/specialist clinics - public and private</td>
<td>Gastroenterologists</td>
<td></td>
<td></td>
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<tr>
<td>Hospital outpatients</td>
<td>Dietitians</td>
<td></td>
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<tr>
<td>Community &amp; child health centres</td>
<td>Physicians</td>
<td></td>
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<tr>
<td>Allied health/dietetic services</td>
<td>Paediatricians</td>
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<td>NGO</td>
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### STAGE 4. BEST LONG TERM CARE AND SUPPORT

<table>
<thead>
<tr>
<th>WHAT</th>
<th>WHO</th>
<th>WHERE</th>
<th>POLICY</th>
</tr>
</thead>
</table>
| ▪ Consistent, best practice care will be provided to individuals diagnosed with coeliac disease | ▪ Organisation: General Practice  
▪ Organisation: Gastroenterology/specialist clinics - public and private  
▪ Organisation: Hospital outpatients  
▪ Organisation: Community & child health centres  
▪ Organisation: Allied health/dietetic services  
▪ Organisation: NGO | ▪ Individual: GPs, practice nurses  
▪ Individual: Gastroenterologists  
▪ Individual: Dietitians  
▪ Individual: Physicians  
▪ Individual: Paediatricians | ▪ General Practice  
▪ Gastroenterology/specialist clinics - public and private  
▪ Hospital outpatients  
▪ Community & child health centres  
▪ Allied health/dietetic services  
▪ NGO | ▪ General Practice  
▪ Gastroenterology  
▪ Allied health/dietetics  
▪ NGO  
▪ Professional associations - ADA, AGA  
▪ DOH/Health Networks |

All individuals diagnosed with coeliac disease in WA will receive periodic assessment for the presence of long-term complications or comorbidities such as osteoporosis. If these are present a management plan will be put in place.

Individuals who develop complications of coeliac disease will receive timely, appropriate management.

All people in WA with coeliac disease will have access to high quality services.