How to Organize the Transition from Paediatric Care to Adult Health Care

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1 Introduction

Celiac disease (CD) is a chronic, multiorgan autoimmune disease that affects the small bowel in genetically predisposed persons precipitated by the ingestion of gluten [1]. CD is one of the most common chronic gastrointestinal diseases [2, 3]. The treatment is primarily a gluten-free diet (GFD), which requires significant patient education, motivation, and follow-up. People with CD should be monitored regularly for residual or new symptoms, adherence to GFD, and assessment for complications. In children, special attention to assure normal growth and development is recommended [4].

The concept of transition consists of a gradual process of empowerment that equips young people with the skills and knowledge necessary to manage their own healthcare in paediatric and adult services. Effective transition has been shown to improve long-term outcomes [5]. The organization of transition is a dynamic process, aiming at ensuring continuity, coordination, flexibility, and sensitivity in a multi-disciplinary context, to meet the adolescent's clinical, psycho-social, and educational needs as well as enhance his/her abilities [6].

For chronic gastrointestinal conditions such as inflammatory bowel disease, CD, and chronic liver diseases with a paediatric onset, patients should undergo a transition process during adolescence. The transition of adolescents from paediatric to adult care is a crucial moment in managing chronic diseases such as CD. The

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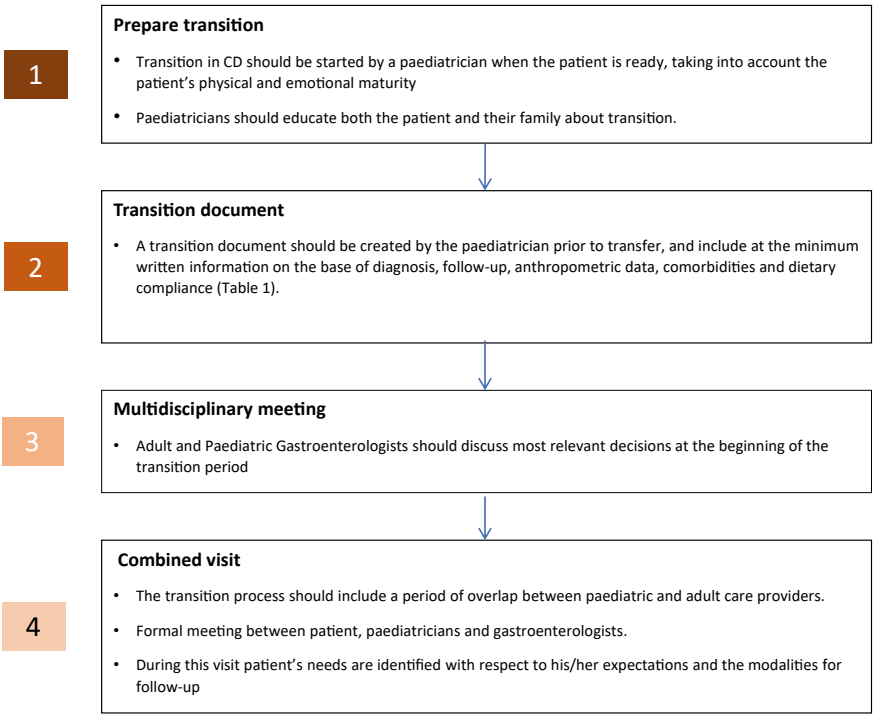

Fig. 1 Transition steps from paediatric to adult care

transition phase for young CD patients is pivotal in maintaining optimal quality of life and a long-term prognosis comparable to the general population (Fig. 1).

2 Specific Aims for Transition

The Prague consensus report [7] proposes recommendations for the management of CD in adolescents and young adults, and how to facilitate the transition to adult healthcare for patients with CD. The transition process should gradually parallel the evolution of child to adult and include an incremental transfer of responsibility for self-care to the adolescent patient with CD. Transition is a complex process, and specific aims in adolescents and young adults are:

- Encourage maturation of communication and decision-making skills.
- Allow patients to take responsibility for medical self-management.
- Education and counselling of the adolescent/young adult to manage a gluten-free diet and consequences of non-adherence.

- Recognition and treatment of psychological problems: discouragement, feeling overwhelmed, anxiety about the future and complications such as depression and eating disorders.
- Increase disease knowledge and its potential complications.
- Help the patient develop good health habits and self-care skills that encourage autonomy and establish good health habits.
- Address the family's anxieties or questions.

3 The Actual Transfer of Care

The process of transition from childhood to adulthood is characterized by physical, mental, and psychosocial development. Data on the transition and transfer of care in adolescents/young adults with CD are scarce.

Generally, paediatric transition to adult care should involve patients, their parents or caregivers, the physician, and the dietician. Although difficult to establish, a position statement [6] by the Italian Societies of Gastroenterology suggests that the ideal age for transition is between 16 and 20 years, depending on physical and emotional maturity, disease activity, adherence to treatment, and autonomy disease management. Thus, paediatricians should decide when their young patients are ready to start the transition programme. In a joint statement [8], three physician organizations suggest that the physician starts a discussion about transition when the adolescent is 12–13 years old and develops a transition plan at 14–15, with the actual transfer taking place at ≥ 18 years of age. Cultural and social differences, as well as individual patient preferences, mean variations may occur.

Ideally, at the beginning of adolescence, the paediatric Gastroenterologist should educate both the patient and their family about transition, make the patient gradually autonomous in managing his/her chronic disease, and prepare him/her for the later transfer to an adult facility. Research should take advantage of new tools to assess transition readiness (as measured by self-management and advocacy skills, rather than chronological age) to determine when a young person may be ready to transfer.

Adolescent patients are often characterized by low adherence to therapy and should be strictly monitored before starting the transition. Poor compliance to GFD among teenagers can negatively affect both quality of life and clinical course. Regarding the psychological aspects involved, children and adolescents with chronic diseases are at greater risk of long-lasting psychological distress than the general patient population, resulting in non-adherence to their treatment and follow-up regimens.

Growth impairment is a known consequence of untreated or undertreated CD [9], though many children with short stature diagnosed with CD in childhood demonstrate good catch-up growth. Untreated CD, or diagnosed after attainment of adult height, usually results in shorter final height than seen in healthy controls.

While the precise pathophysiology may be poorly understood, some adolescents and young adults with CD will experience a delay in pubertal development. When the transition is anticipated, the paediatrician should provide data regarding the patient's history of physical development and should note to the adult physician whether the patient has achieved his/her final adult height. For those patients who have experienced significant pubertal delay the paediatrician may be better suited to provide guidance and coordinate the transition to adult care after puberty, particularly if other paediatric specialists, as endocrinologists try to manage growth failure. A bone age X-ray may inform growth expectations and timing of transition [7].

Implementing a systematic transition policy in CD has been limited by a lack of clinical guidelines based on outcome-related research. In the absence of solid evidence, different models of transition will likely be developed locally. The actual transfer can take many forms. In some settings, the paediatric and adult gastroenterologists see the patient simultaneously; in others, paediatric and adult gastroenterologists meet annually to discuss patients in transition. Optimally, joint transition clinics with paediatric and adult service clinicians can be established for information delivery and generating trust in the new physician. The Prague consensus [7] recommends that the actual transfer from paediatric to adult care should be structured and include the minimum written information based on diagnosis, follow-up, anthropometric data, comorbidities, and dietary compliance.

These patients may have difficulty communicating with health providers for many reasons about communicating with adolescents and young adults. The presence of a parent can be helpful if the adolescent has not been prepared for independent visits. Young patients may have difficulty expressing sensitive concerns in person to a provider. Still, they may do so more readily by different types of electronic communication, including email, videoconferencing, SMS messaging, and online consultations. This has also been tested in paediatric groups with some success [10].

During the transition period, several issues may be discussed [7].

(1) Some adolescents/young adults may question their diagnosis and feel the transition period is a natural point for discussing how the diagnosis was made and whether re-evaluation is appropriate.
(2) In adolescence, patients with CD should gradually assume the exclusive responsibility for their care, although parental support is still important. The responsibility of keeping a GFD must be shared by the patient and his/her parents.
(3) Adolescence is recognized to be a period when adherence is poor, and these patients report lower adherence than younger children. Therefore, dietary adherence, consequences of non-adherence, and complications despite being asymptomatic are key components for discussion in a transition setting.
(4) Dietary non-adherence in adolescents is associated with increased disease burden, poorer quality of life, and increased physical symptoms. Moreover, patients should know that dietary adherence is essential before conception and during pregnancy as women with untreated CD are more likely to suffer an adverse pregnancy outcome.

Another issue that needs to be discussed is medical monitoring with laboratory tests and healthcare visits. Otherwise, allocating time and space to discuss with experts about psychological aspects could be necessary.

4 Factors Affecting the Transition

The transition of adolescents from paediatric to adult care is crucial in managing chronic diseases such as CD. A smooth transition may encounter obstacles linked to the experience of the patients and their families (caregivers) and the paediatric and adult health care providers. The most effective way to achieve a smooth transition has become a subject of considerable debate. A planned and organized transition of care for adolescents with CD is recommended, though little data are available regarding factors associated with successful transition [11].

The crucial issue of switching from a family-centred (paediatric) care model, with parents' direct involvement in the diagnostic and therapeutic decision-making process, to self-managed (adult) care, may cause a young patient to experience a sense of exclusion and fear. Some barriers to a successful transfer include:

- Lack of coordination between adult and paediatric services,
- Lack of planning and resistance of patient and families to the transition of their healthcare.

An inappropriate transition or the incomplete transmission of data from the paediatrician to the adult gastroenterologist can decrease compliance to a young patient's treatment and prognosis. Otherwise, lack of regular follow-up seems to be a particular problem for the phase of transition between paediatric and adult care [7].

A recent study [12] provides new insights in the transition of care of young adults with CD: patients diagnosed younger show poorer transition rates, and those lacking symptoms are less likely to transition to adult care. Moreover, this study suggests that lifelong adherence to a GFD may differ depending upon age of diagnosis. Individuals with CD diagnosed early in childhood have demonstrated better dietary adherence than individuals diagnosed as older children or adults. Those diagnosed in adolescence may be less adherent than their younger and older comparators.

Transitional periods, such as starting school, have been associated with diminished dietary adherence. Nevertheless, living circumstances (living with a parent or relatives living independently) did not impact the likelihood of transition. Similarly, the level of education attained, when controlling for age, did not influence follow-up. However, another study [13] suggests that compliance and quality of life improve with a better knowledge of the disease. Patients diagnosed later in life might better follow the GFD independently of their knowledge. Moreover, this study reports that self-management and knowledge improved as age increased. A direct correlation between age at diagnosis and dietary adherence is in line with a previous study [14]. Younger age at diagnosis and current age were related to dietary non-adherence. Those currently in their teens were likely to be non-adherent.

A planned and organized transition of care for adolescents with CD is recommended, though little data are available regarding factors associated with a successful transition. Table 1 shows the document used in our institution to obtain relevant information about the child who is going to be transferred to an adult unit. Further studies are needed to identify and remove barriers to transition.

5 Use of Biopsy, CD Serology and Genetic Testing in Transition to Adulthood

European Society of Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) 2012 guidelines [15] suggested, for the first time, that the diagnosis of CD can be made without biopsies in a subgroup of paediatric patients.

New ESPGHAN 2020 guidelines [16] support that the no-biopsy approach for CD diagnosis is confirmed to be safe in children with high TGA IgA values ≥ 10 times the upper limit of normal with accurate, appropriate tests and positive endomysial antibodies (EMA IgA) in a second serum sample. The updated review of the 2012 criteria in 2020 provide new evidence on some aspects, such as the role of HLA and the diagnosis of asymptomatic patients. This test would only be indicated for screening of at-risk individuals and in case of uncertain diagnosis. On the other hand, this guideline gives a conditional recommendation that, taking available evidence into account, CD can be diagnosed without duodenal biopsies in asymptomatic children, using the same criteria as in patients with symptoms.

Therefore, the are some differences in the use of histology for diagnostic purposes in children and adults:

- One is the no-biopsy approach in children, in selected cases; while in adults, guidelines [4, 17] emphasize the combined use of biopsy and serological analyses for diagnosis.
- Other difference is that normal architecture with increased intraephitelial lymphocytes (IELs) is considered non-specific in paediatric guidelines whereas IELs $\geq 25/100$ enterocytes have been validated as a cut-off point in adults.
- In children, Marsh 1 is not considered sufficient to diagnose CD, but some observations suggest that potential CD cases with Marsh 1 small bowel lesions have a higher chance to evolve to villous atrophy in comparison to Marsh 0. Patients with no/mild histological changes (Marsh 0/I) but confirmed autoimmunity (TGA IgA/EMA–IgA+) should be followed closely.

The no biopsy policy adopted by ESPGHAN guidelines may present a topic for discussion in paediatric to adult care. If the patient was diagnosed according to the ESPGHAN criteria is necessary to review the symptoms, results of serology, HLA status and response to GFD.

If the existing diagnostic guidelines have not been met, and the diagnosis needs re-evaluation, a new diagnostic approach should be instituted. Serology and

Table 1 Items to be included in transition document that should accompany the adolescent/young adult to adult healthcare in CD

Name
Date of birth
Diagnosis of celiac disease, year, and name of the Institution
Has the patient (or their relatives) been registered with an official patient association?
Weight and height at the time of transition (BMI)
Presentation pattern (e.g., anaemia, growth retardation, malabsorptive diarrhoea)
History of fractures (YES/NO) If so, specify
Age at the time of menarche
Serology at diagnosis (please indicate the value with range of normality)
Histology at diagnosis (please indicate grade of lesions)
HLA status if available
Associated diseases (thyroid diabetes, other)
Clinical response to gluten-free diet • Symptomatic response (YES/NO) • Histological response (if available) (None, partial, total)
Is there an associated intestinal condition as a cause of "*unresponsive celiac disease*"? – Sugar intolerance – Intestinal bacterial overgrowth – Pancreatic exocrine insufficiency – Microscopic colitis – Irritable bowel syndrome – Crohn's disease – Giardiasis – Others • Should any relevant psychosocial factor be named? (YES/NO) If so, Specify • Is there any identifiable psychiatric comorbidity? (YES/NO) If so, Specify

CD: celiac disease; BMI: Body mass index; HLA: human leukocyte antigen

histology may be part of this approach. In adolescents and young adults, biopsy to reconfirm a childhood diagnosis of CD may be considered when the tenfold positive TGA-IgA result has not been confirmed by positive EMA in a second serology at the time of diagnosis or when the ESPGHAN diagnostic criteria have not been met in a child without duodenal biopsies. Biopsies may also be relevant when the adolescent has ceased a GFD because he or she doubts the diagnosis, the patient or the physician requires documentation of healing, and the presence of symptoms suggests active CD. A gluten challenge is indicated before the biopsy. Moreover, HLA testing can be used to rule out CD in unclear cases. As the adult patient depends on his/her own judgement to follow dietary instructions it is strongly recommended that a definite diagnostic decision, based on the above-mentioned criteria is established before transition. If diagnosis is in doubt or there was inconsistent protocol, compliance may be questioned.

It is also important that the paediatric and adult physicians agree on the same criteria to avoid confusing the patient or questioning the real need for the GFD.

6 Follow up

Follow-up of patients with CD is recommended to ensure dietary adherence, prevent, or detect complications or associated conditions, including autoimmune thyroid disease, and promote optimal health. Data suggest continued follow-up improves dietary adherence. Based on expert opinion, all paediatric patients should be seen at 3–6 months intervals for the first year after diagnosis. Once symptoms have resolved and serological tests for CD have normalized, an annual follow-up visit is recommended.

CD is associated with fracture risk [18], predominantly before treatment or in the setting of non-adherence to GFD. Bone mineral density is frequently depressed in both children and adults with CD at the time of diagnosis, and deficits have been shown to correlate with the degree of histological severity. Most children recover from bone mineral density abnormalities following appropriate therapy. Thus, dual-energy X-ray absorptiometry should only be considered for young adults at high risk.

7 Primary Care Involvement

In many countries, adolescents leaving paediatric care are often cared for by a general practitioner rather than by an adult gastroenterologist. Primary care physicians (PCPs) are then also responsible for the healthcare during and after transition. In adults, PCPs may take a major role in care. Some adolescent/young adult patients are also referred to primary care when they are considered healthy after diagnostic workup information and initial follow-up in secondary care (either

with a paediatrician or an adult gastroenterologist). Primary care may be a suitable care provider if adequate personnel skills and laboratory facilities are sufficient for long-term follow-up, and this may depend on local practice. In the authors' opinion, a joint follow-up by both bodies (PCPs and gastroenterologists) will be necessary in many cases.

8 Conclusions

The transition between paediatric and adult care for young people is now recognized as a key component of care, across the spectrum of physical and mental illness and disability, though there has been little high-quality evaluation published. Transferring care in an organized manner has been associated with improved outcomes, such as a greater feeling of preparedness in young patients with chronic illness and improved adherence with medical care. The transition team has the delicate task of assisting young adults and their families in understanding and appreciating the cultural and practical differences between paediatric and adult medicine. An effective transition can avoid gaps in medical care and ensure physical and mental well-being during this difficult time.

In the absence of solid evidence, different models of transition could vary both nationally and internally. Models of transition will eventually need to be evaluated in randomized controlled trials with clear patient outcome measures. Socio-economic effectiveness and outcomes of care of the different models should also be carefully evaluated.

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