Nutritional Management In Cystic Fibrosis

Transition Of Cystic Fibrosis Patients From Childhood To Adulthood

F. Huet (FR)
Transition of CF patients from Childhood to Adulthood

Frédéric HUET, MD, PhD
DIJON - FRANCE
• No Conflict of interest in this medical field
CF epidemiology: a new paradigm

A Stephenson et al. Presse Med 2017; 46:e87-e95
Life expectancy and survival charts

A Stephenson et al. Presse Med 2017; 46:e87-e95
CF epidemiology

- More than 80 000 CF patients worldwide
- Predicted life expectancy > 45 years in western countries

⇒ **Not a lethal pediatric disease but a life-span shortening multiorgan chronic condition in adults**

⇒ **Development of transition programs to facilitate transfer of care from pediatric to adult care providers**
Definition

• **Transfer**: actual point in time when the patient moves from pediatric to adult care providers.

• **Transition**: purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care programs.

• **Process requiring planning**

Danger of sudden transfer +++
Gaps in care during transfer

Median gap: 106 days

Modeling the probability gap > 365 days:
- age of last visit < 18
- different city
- no health insurance

Gender and FEV1% are not significant predictors

Risks of an inappropriate transfer

- Long gap in follow-up
- Large differences in the therapeutic protocol
- Lack of confidence in care workers and medical teams
- A sudden personal commitment to forego previous habits is required
- Multiplication of interlocutors (bone, diabetes, liver etc.) justifying the expertise of a multidisciplinary team and not punctual disciplinary individual experts
- ....
What is the appropriate moment for transition?

From CFF Patients Registry 2014 Annual Data Report. Bethesda, Maryland
Height velocity in boys during puberty

Contribution of puberty on final height = 15.53%

Nutrition?

Huet F et al Arch Dis Child 2012
Age-appropriated transition goals for patients with CF

<table>
<thead>
<tr>
<th>12 y</th>
<th>13–14 y</th>
<th>15–16 y</th>
<th>17–18 y</th>
<th>19–21 y</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Transition goals</strong></td>
<td>Describe what CF is</td>
<td>List medications, amounts, and times when taken</td>
<td>Become better at recognizing symptoms and describing them</td>
<td>Contact CF caregivers directly to discuss changes in health</td>
</tr>
<tr>
<td><strong>Name medications and reasons for taking them</strong></td>
<td>Answer questions independently in clinic</td>
<td>Describe choices about smoking and drinking and effect on health</td>
<td>Schedule appointments and tests</td>
<td></td>
</tr>
<tr>
<td><strong>Take enzymes</strong></td>
<td>Recognize changes in symptoms and describe them</td>
<td>Be aware of clinic and test appointment dates</td>
<td>Refill prescriptions</td>
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<tr>
<td><strong>Remember to do airway clearance</strong></td>
<td>Do airway clearance without help</td>
<td>Be aware of medication supply and need for refills</td>
<td>Maintaining own equipment</td>
<td>Learn details about insurance coverage</td>
</tr>
</tbody>
</table>

Data from Boyle MP. Transitioning to adult care: a transition for parents as well! Article on MyCysticFibrosis.com web site 2007
Clinical effects of transition in CF patients

Retrospective 1:1 matched cohort study from the CFF Patient Registry. Comparison of 661 «transfer-positive» patients with 661 patients with similar background characteristics cared for in pediatric CF programs.

<table>
<thead>
<tr>
<th>Outcome Measure</th>
<th>Estimate (95% CI)</th>
<th>P Value</th>
</tr>
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<tbody>
<tr>
<td>Percent predicted FEV₁</td>
<td>0.76 (−0.30 to 1.83)</td>
<td>.159</td>
</tr>
<tr>
<td>Percent predicted FVC</td>
<td>1.40 (0.40 to 2.40)</td>
<td>.006</td>
</tr>
<tr>
<td>BMI</td>
<td>0.22 (0.04 to 0.39)</td>
<td>.015</td>
</tr>
<tr>
<td>Number of hospitalizations</td>
<td>−0.04 (−0.15 to 0.08)</td>
<td>.535</td>
</tr>
<tr>
<td>Number of home IV antibiotic events</td>
<td>0.08 (−0.03 to 0.19)</td>
<td>.134</td>
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=> No significant alteration of short-term outcomes among youth who remain in care

Transition and nutritional challenges (1)

• **Prevention of malnutrition.** Patients who have had long-term experience of caloric intakes and supplements.

• **Detection and management of CF Related Diabetes.** Early detection and treatment of glucose intolerance ++++. Major negative impact of CFRD on pulmonary status. Opposition between dietary restrictions of type 1 and 2 diabetes and requirements of CFRD *(high-fat, high-energy, high-protein, high-Na diet).*

• **Bone health.** Multiple factors explain low bone mineral density in CF (CFTR mutations, poor nutritional status, deficit of the fat-soluble vitamins, poor Ca intakes, recurrent chest infections, delayed puberty, CFRD, reduced physical exercise, corticoids use...).
Transition and nutritional challenges (2)

- **Social pressure.** Standards of beauty revolving around being thin and eating « healthy » low-fat high-fiber diets.

- **Eating disorders and its behaviors.** Females with CF are usually skinny and are happy with their perceived body image. **Goal : BMI ≥ 22 kg/m²**

- **Treatment of denutrition.**
Challenges of the Nutritional Transition

• If nutritional counselling or oral supplements fail to prevent or reverse poor nutrition => enteral tube feeding

• But additional factors may be taken into consideration:

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Burden of treatment and transition

• Most young adults: regimen associating
  • Pancreatic enzyme-replacement therapy at every meal and snack
  • Supplemental vitamins and hypercaloric diet
  • Prophylactic oral antibiotic, inhaled and nebulised therapies
  • Physiotherapy

Large number and complexity of these treatments

• More complex care
  • Liver disease
  • CF related diabetes
  • CF related low bone mineral density
  • O₂ therapy and/or non-invasive ventilation
  • IV antibiotic therapy
  • Enteral tube feeding

=> Understanding
=> Improving adherence ++++
Adherence of CF patients to their Home Programs

• Transition = autonomous management and adherence of treatment programs

• Evaluation of this adherence?
  ➢ Pancreatic enzyme replacement therapy (9 studies): mean adherence between 27.4 to 96.5 %
  ➢ Calorie intake (10 studies): reported adherence between 40.8 and 81 %
  ➢ Vitamine intake (8 studies): mean adherence from 22 to 97.5 % (disparity due to variability of vitamins)
  ➢ Dietitian’s food guidelines (1 study): mean adherence from 70% (patients) to 80% (parents)

O’Donohoe et al Respiratory Care 2014; 59:1731
Conclusion

• From Childhood to Adulthood: a big and stimulating challenge!

• Transition needs anticipation, education, communication (patient and parents) and an expert multidisciplinary team

• Nutrition is one of the hot spot of this transition in CF
L’équipe pluridisciplinaire

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